Creutzfeldt-Jakob disease: Magnetic Resonance Imaging Findings

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Purpose

To describe the magnetic resonance imaging (MRI) appearance of Creutzfeldt- Jakob disease (CJD) and evolution of lesions.

Methods and Materials

We retrospectively reviewed clinical notes, conventional magnetic resonance (MR) sequences and diffusion-weighted imaging (DWI) in four patients with CJD.

The four patients were male with an age range between 53 and 62 years old and a mean age of 57 years.

The patient data and the MRI were collected in 2001-2009.

The first MR examination was performed within 15 days, 1, 2 and 4 months of onset of symptoms and the second MR examination was performed 10, 15, 24 and 49 days later.

Six examinations were performed with a 1, 5 T unit and two with a 3 T unit.

Results

Patients had striatal lesions or cerebral cortical lesions or both, in the early phase of the disease.

Case 1:

1st MR exam shows hyperintense lesions at T2-weighted images, FLAIR images and DWI in basal ganglia and in the cingulated cortex.

2nd MR exam: left hippocampus appears hyperintense at FLAIR images, and high signal in the globus pallidus and thalamus on T1-weighted images.

Case 2:

1st MR exam: hyperintense lesions in cortex cerebral at FLAIR images and DWI with normal basal ganglia.
2\textsuperscript{nd} MR exam: hyperintense lesions in caudate and left putamen.

**Case 3:**

Hyperintense lesions in cortex cerebral at FLAIR and DWI with normal basal ganglia in both examinations.

**Case 4:**

1\textsuperscript{st} MR exam: hyperintense lesions in right caudate, putamen and frontal, parietal and occipital cerebral cortex.

2\textsuperscript{nd} MR exam: hyperintense lesion in left caudate.

<table>
<thead>
<tr>
<th>Nº Patient</th>
<th>Clinical Findings in CSF</th>
<th>Duratio\textsuperscript{M. R.} of CJD (months)</th>
<th>Polymorphic Codón 129</th>
<th>Diagnostic level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>age</td>
<td></td>
<td>Imaging findings</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>male</td>
<td>Ataxia + memory loss.</td>
<td>6</td>
<td>Homozygous MM</td>
</tr>
<tr>
<td></td>
<td>53</td>
<td></td>
<td></td>
<td>Confirmed at histopathologic analysis</td>
</tr>
<tr>
<td>2</td>
<td>male</td>
<td>Gait + disturbances, disarthria, visual disturbances and optical hallucinations</td>
<td>3</td>
<td>Not done</td>
</tr>
<tr>
<td></td>
<td>62</td>
<td></td>
<td></td>
<td>Sporadic CJD</td>
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</table>
Case 1: sporadic CJD in a 53-year-old man with ataxia.

1st MR exam: 4 months after onset of symptoms.

Axial T2-weighted images (Figures 1 and 2) and coronal FLAIR image (Figure 3) show abnormal high signal intensities in basal ganglia: caudate nuclei and putamen.

Axial DWI images (Figures 4 and 5) show high signal intensities in bilateral basal ganglia.

2nd MR exam: 24 days after the 1st MR exam.

Axial T2-weighted fast spin-echo images (Figures 6 and 7) show high signal in basal ganglia.

Coronal FLAIR images (Figures 8 and 9) show high signal in basal ganglia and left hippocampus.

Axial FSE T1-weighted image (Figure 10) and Axial FSE T1-weighted images with magnetization transfer prepulse (Figures 11 and 12) show high signal in globus pallidus. Histopathological findings indicate an exceptionally heavy deposition of prion protein in this area and it seems likely that this caused the high signal in the media part of the globus pallidus. Although the putamen had ever higher protein content, the T1-shortening effects of prion protein in this area are probably cancelled out by the coexistent high degree of...
spongiform degeneration, leading to an overall T1 relaxation time longer than that of the globus pallidus.

The patient died 6 months after onset of symptoms.

Cerebrospinal fluid examination for the 14-3-3 protein was positive. Gene mutation analysis at codon 129 showed homozygosity for methionine.

Brain autopsy was performed and demonstrated diffuse gray matter spongiform degeneration, which confirmed the diagnosis of sporadic CJD.

**Case 2**: A 62-year-old man with a 2-week history of weakness, unsteady gait, dysarthria, visual disturbances and involuntary movements of the left arm.

**1st MR exam at 3 T unit**: 15 days after onset of symptoms.

Coronal Flair images (Figs. 13, 14, 15 y 16) show abnormal high signal intensities in the occipital and parietal cortex bilaterally with normal signal in the basal ganglia.

Axial FLAIR images (figs. 17, 18, 19 and 20) show abnormal high signal intensities in the parieto-occipital cerebral cortex bilaterally.

Differential diagnosis of early-stage CJD with mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS), viral encephalitis and hypoxia. In these cases lesions show severe gyriform swelling and contrast enhancement on T1-weighted images, which does not extend to the underlying white matter. The affected regions in herpes simplex encephalitis show necrotic or hemorrhagic areas. These features are never seen in early-stage of CJD.

In early-stage CJD, high-signal-intensity abnormalities are most frequently observed in the cortex. The cortical abnormalities may be unilateral, bilateral, diffuse or focal, and symmetric or asymmetric. Abnormalities of the basal ganglia also may be unilateral or bilateral, and the caudate is most often involved. The high-signal-intensity lesions depicted on DWI are thought to represent vacuolization of neuropil.

Abnormal DWI changes may be present in patients with CJD who have normal EEGs and normal CSF studies.

During the intermediate phase, the area of high signal intensities on the DWI had expanded from the caudate nucleus to involve the putamen.
During the terminal phase abnormal high signal intensities in the cerebral cortex and basal ganglia on the DWI can disappear.

Axial DWI (figs. 21, 22, 23, 24 and 25) reveal abnormal high signal intensities in parieto-occipital cerebral cortex bilaterally and in the left temporal cerebral cortex.

The reason for cortical signal intensity changes in CJD is not fully understood. Several studies have shown that the intensity of MR changes correlates with the neuropathologic findings: spongiosis, neuronal loss and astrogliosis. In areas that are bright on MRI in FLAIR or DWI, neuropathologic changes are more severe than in areas with normal cortical signal intensity.

The cortical signal intensity changes correlate with clinical signs. In this patient the affection of the occipital cortex explain the visual disturbances.

2nd MR exam (1.5T): 49 days after the 1st MR exam. The patient’s neurological state rapidly deteriorated with optical hallucinations, myoclonus and personality changes with aggressiveness and agitation.

Coronal FLAIR images (figs. 26 and 27) show areas of abnormal high signal intensity in the parieto-occipital cerebral cortex.

Axial DWI (fig. 28) reveals not only cortical lesions but also diffuse bilateral striatal lesions.

Axial ADC map from DWI (Fig. 29) shows the lesions as areas of decreased signal intensity, suggesting the presence of restricted diffusion within the tissue. Bahn and Parchi correlated abnormal DWI findings with vacuole formation in the brain.

CSF examination for the 14-3-3 protein was positive.

The patient died three months after the onset of symptoms.

Case 3: A 57-year-old man with rapidly progressive dementia with visual disturbances, apraxia and acoustic and optical hallucinations.

1st MR exam: 1 month from the onset of symptoms.
Coronal FLAIR images (figs. 30 and 31) reveal only subtle hypertense lesions in the occipital cerebral cortex bilaterally, without lesions in the basal ganglia.

2\textsuperscript{nd} MR exam: 15 days from the 1\textsuperscript{st} MR exam.

Coronal FLAIR images (Figs. 32 and 33) show high signal intensities in occipital cerebral cortex bilaterally, now appear more prominent.

Abnormal signal hyperintensities of the brain cortex without changes in the basal ganglia or thalami have been reported in only a few patients, and may reflect the early stage of the disease or perhaps phenotypic or genotypic CJD variants.

CSF examination for the 14-3-3 protein was positive.

The patient died at fifth month from the onset of the symptoms.

**Case 4:** A 56-year-old man had a two month history of cerebellar signs, double vision, memory loss, lack of fluent speech and irritability.

1\textsuperscript{st} MR Exam obtained two months after the onset of symptoms.

Axial FLAIR images (figs. 34, 35, and 36) reveal bilateral areas of abnormal high signal intensity in the fronto-parieto-occipital region of the cerebral cortex and right basal ganglia.

2\textsuperscript{nd} MR Exam (3T) obtained 10 days after the 1\textsuperscript{st} MR Exam.

Axial FLAIR images (figs. 37 and 38) and Axial T2 image (fig. 39) show high signal intensity in the cerebral cortex and in the basal ganglia.

Axial DWI (figs. 40, 41 and 42) reveal high signal intensity in the fronto-parieto-occipito-temporal cerebral cortex bilaterally and in the basal ganglia.

Axial ADC map from DWI (fig. 43) shows the basal ganglia lesions as areas of decreased signal intensity.
DWI has a higher conspicuousness for the detection of cortical changes compared with FLAIR. The DWI allows easiest identification of the signal intensity changes and should therefore be included in the work-up of patients with suspected CJD.

CSF examination for the 14-3-3 protein was positive.

The patient died 3 months after the onset of the disease.

The differential diagnosis of CJD should be clinically differentiated from other disorders associated with dementia as Alzheimer disease, which is not characterized by abnormalities on DWI.

Vascular dementia is associated with evidence of multiple infarcts, but DWI abnormalities are observed only in the area of a recent infarction, and there is no diffuse cortical involvement.

MELAS is a genetic metabolic disorder that induces subacute dementia, mitochondrial myopathy, encephalopathy, lactic acidosis and strokelike episodes. Although MELAS may mimic CJD, it typically is manifested in a younger age group than is CJD.

On DWI abnormal cortical signal hyperintensities are depicted in the early stage of MELAS. On T2-weighted MRI and FLAIR images the lesions appear as gyriform areas of severe swelling that spares the underlying white matter, a feature that is never observed in early-stage CJD.

Venous hypertensive encephalopathy: several disease processes may cause venous hypertensive encephalopathy. Dural arteriovenous fistula has received attention as a potential cause of reversible dementia.

On DWI, venous infarction sometimes is depicted as a high-signal-intensity abnormality in the cortex that resembles a lesion in CJD.

Three-dimensional gadolinium-enhanced MR angiography may help achieve the correct diagnosis.

Chronic herpes encephalitis: Herpes simplex encephalitis is the most common viral infection of the central nervous system. On T2 weighted MRI, high signal-intensity abnormalities typically are seen in the medial temporal regions and in some cases involve the frontal lobe. The affected regions in herpes simplex encephalitis show necrotic or hemorrhagic parenchymal swelling in the early stage, a feature that is never observed in CJD.
Images for this section:

Fig. 1: axial T2-weighted image
Fig. 2: axial T2-weighted image
Fig. 3: Coronal FLAIR image
Fig. 4: axial DWI
Fig. 5: axial DWI
Fig. 6: axial T2-weighted image
Fig. 7: axial T2-weighted image
**Fig. 8:** Coronal FLAIR image
**Fig. 9**: coronal FLAIR image
Fig. 10: axial FSE T1-weighted image
**Fig. 11:** axial T1 with magnetisation-transfer prepulse.
Fig. 12: axial T1 with magnetisation-transfer prepulse.
Fig. 13: coronal FLAIR image
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Fig. 15: Coronal FLAIR image
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Fig. 17: axial FLAIR image
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Fig. 26: coronal FLAIR image
Fig. 27: coronal FLAIR image
Fig. 28: axial DWI
Fig. 29: axial ADC map from DWI
Fig. 30: coronal FLAIR image
Fig. 31: coronal FLAIR image
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Fig. 33: coronal FLAIR image
Fig. 35: axial FLAIR image
**Fig. 36:** axial FLAIR image

![Axial FLAIR Image](image)

**Fig. 37:** axial FLAIR image
Fig. 38: axial FLAIR image
Fig. 39: axial T2-weighted image
Fig. 40: axial DWI
Fig. 41: axial DWI
Fig. 42: axial DWI
Fig. 43: axial ADC map from DWI
Conclusion

CJD should be suspected in any case in which areas of abnormal signal hyperintensities are depicted on FLAIR and DWI in the cerebral cortex and deep gray matter, especially in the caudate nucleus.

Noninvasive early diagnosis helps to prevent the transmission of CJD:

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