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B C Tzeng, C Y Chen, C C Lee, F H Chen, T Y Chou and R
A Zimmerman

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Rapid Spongiform Degeneration of the Cerebrum and Cerebellum in Creutzfeldt-Jakob Encephalitis: Serial MR Findings

Bing-Chang Tzeng, Cheng-Yu Chen, Chau-Chin Lee, Fu-Hwa Chen, Ting-Ywan Chou, and Robert A. Zimmerman

Summary: We report the cerebral MR imaging findings in a patient with pathologically proved Creutzfeldt-Jakob disease in whom predominant gray and white matter degeneration was seen within 1 year of symptom onset. The initial MR signal abnormalities in the basal ganglia were subtle. A follow-up MR examination revealed diffuse cerebral and cerebellar atrophy and demyelination.

Index terms: Dementia; Encephalitis

Creutzfeldt-Jakob disease (CJD), a progressive, inevitably fatal illness, is difficult to distinguish clinically from other forms of dementia. Several case reports have described hyperintense signal abnormalities in the caudate nucleus, globus pallidus, and putamen on T2-weighted magnetic resonance (MR) images (1, 2). Abnormalities of gray and white matter have also been reported (3–6). We describe a pathologically proved case of CJD in which MR findings of global cortical atrophy of the cerebrum and cerebellum were accompanied by white matter degeneration shortly after onset of initial symptoms.

Case Report

A 48-year-old woman was admitted to our hospital with a 1-month history of lethargy, loss of appetite, and poor mental performance. On admission, she exhibited inattentiveness, lack of fluent speech, tremor, and dystonic movement of the right hand. Her medical and family history were unremarkable. Neurologic examination showed her to be alert but with mutism. Her mental performance was characterized by poor calculative abilities and sluggish judgment as to person and place. Muscle strength was normal, but with mild axial rigidity. The deep tendon reflexes were increased, but the Babinski sign was not present. A wide-based, shuffling gait was observed. Cerebrospinal fluid analysis was normal. An electroencephalo-

gram (EEG) showed a generalized pattern of theta-delta background slowing activity and periodic synchronized sharp 1/s delta-wave discharge. Findings at cranial computed tomography (CT) were unremarkable (Fig 1A). The initial impression was dementia of unknown origin.

Two months later, the patient's condition had deteriorated to global aphasia and she was bedridden. Myoclonic jerk was present and muscular rigidity developed in all limbs. There was muscle wasting, followed by difficulty in swallowing and incontinence of urination. Initial, noncontrast MR images revealed a subtle increase in signal in the heads of the caudate nuclei, putamina, and thalami on T2-weighted images (Fig 1B–E). An open brain biopsy of the right temporal lobe performed just after MR imaging revealed severe loss of neurons, hypertrophic glial reaction, and vacuolar-spongiform changes in the gray matter (Fig 1F).

One year later, the patient was in a vegetative state with poor respiratory control. Noncontrast T2-weighted MR images obtained at this time revealed severe cerebral and cerebellar atrophy, prominent dilatation of the ventricular system, and a global increase in signal intensity in the white matter with shrinkage of the basal ganglia (Fig 1G–J).

One and a half years after admission, she was discharged to a chronic-care unit, where she remained in a vegetative state. She died 8 months later.

Discussion

In the past, CJD was considered a slowly developing viral disease because of its prolonged latency followed by a progressively downhill clinical course. It is now thought to be a chronic infectious disease caused by an agent called a *prion*. The prions and viruses have many differences in their properties, structure, and methods of replication. Prions contain little or no nucleic acid and evoke no immune response (7, 8).

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From the Department of Diagnostic Radiology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, Republic of China (B.-C.T., C.-Y.C., C.-C.L., F.-H.C., T.-Y.C.); and the Department of Radiology, the Children's Hospital of Philadelphia (Pa) (R.A.Z.).

Address reprint requests to Ting-Ywan Chou, MD, Tri-Service General Hospital, National Defense Medical Center, 8, Sec 3, Ting-Chou Rd, Taipei, Taiwan, Republic of China.

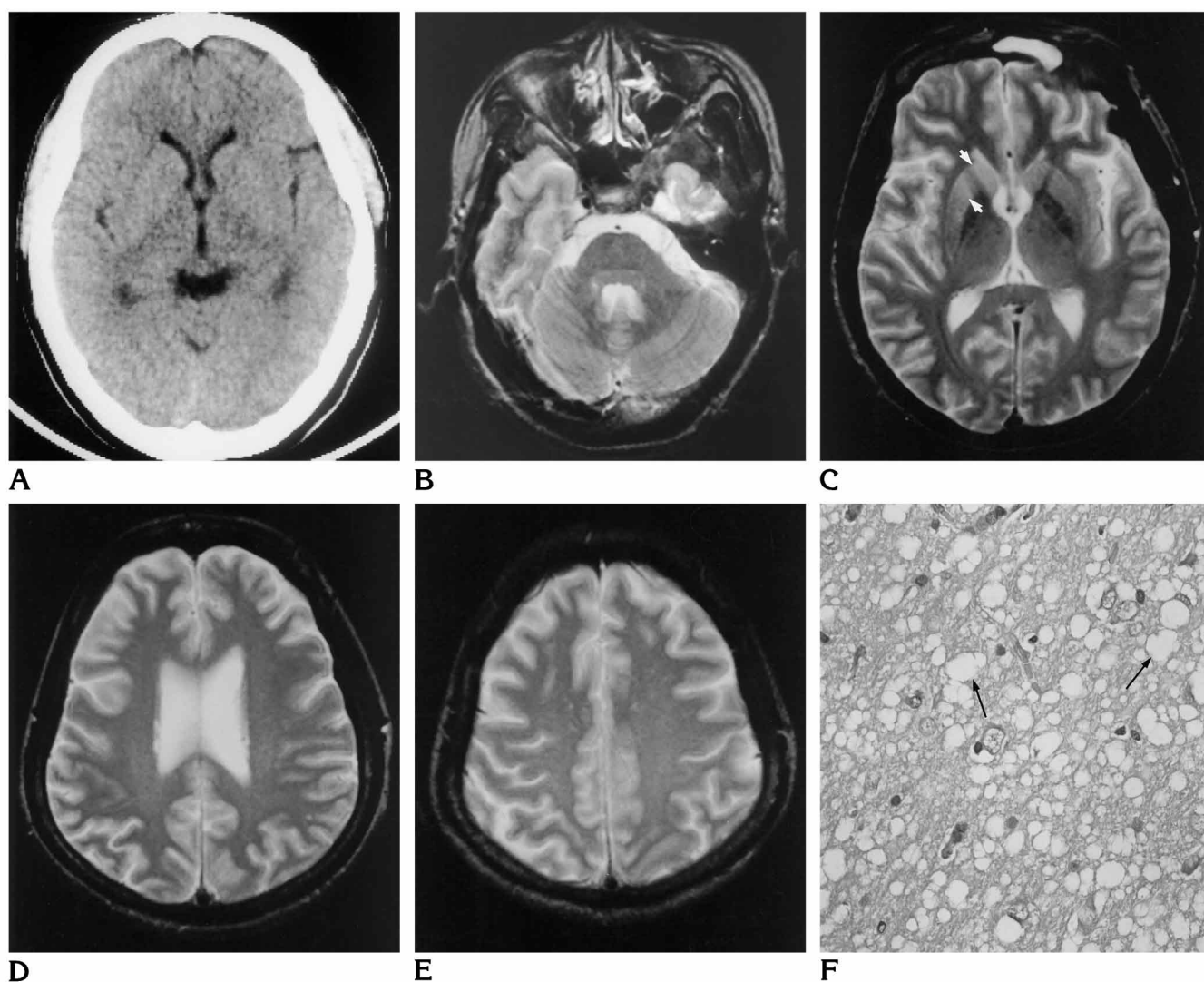


Fig 1. A, Initial noncontrast CT scan at the level of the basal ganglia is unremarkable.

B–E, T2-weighted MR images (2500/90/1 [repetition time/echo time/excitations]) at the midpons (B), basal ganglia (C), body of the lateral ventricles (D), and centrum semiovale (E) obtained 2 months after A show a mild increase in signal intensity of the right corpus striatum (arrows in C).

F, Histopathologic section of the surgical biopsy specimen obtained from the right temporal gray matter shows neuronal loss and fine vacuolation and spongiform change (arrows) (hematoxylin-eosin, ×400). *Figure continues.*

CJD is a progressive, inevitably fatal disease of the central nervous system with a mean survival time of 6 months to 1 year after diagnosis. The disease is transmissible to a variety of species, and human-to-human iatrogenic transmission has been well documented. CJD should be considered in patients who have a rapidly progressive dementia that appears in mid-adult life, especially if accompanied by myoclonic seizures, as was seen in our case. There are no reliable tests for the early diagnosis of the disease.

The typical EEG pattern is periodic high-volt-

age sharp waves against a background of low-voltage activity. This pattern, however, is not always present and is nonspecific (9). CT scans usually show either no abnormalities (80% of cases) or nonspecific atrophy (10). Metabolite alterations detectable by MR spectroscopy are not sufficiently established to allow an early diagnosis (11). Even now, the diagnosis can only be made by brain biopsy. Typical pathologic changes in the involved brain may show spongiform degeneration with prominent depletion of neurons and astrocytes and marked gliosis in the cortex and white matter (1).

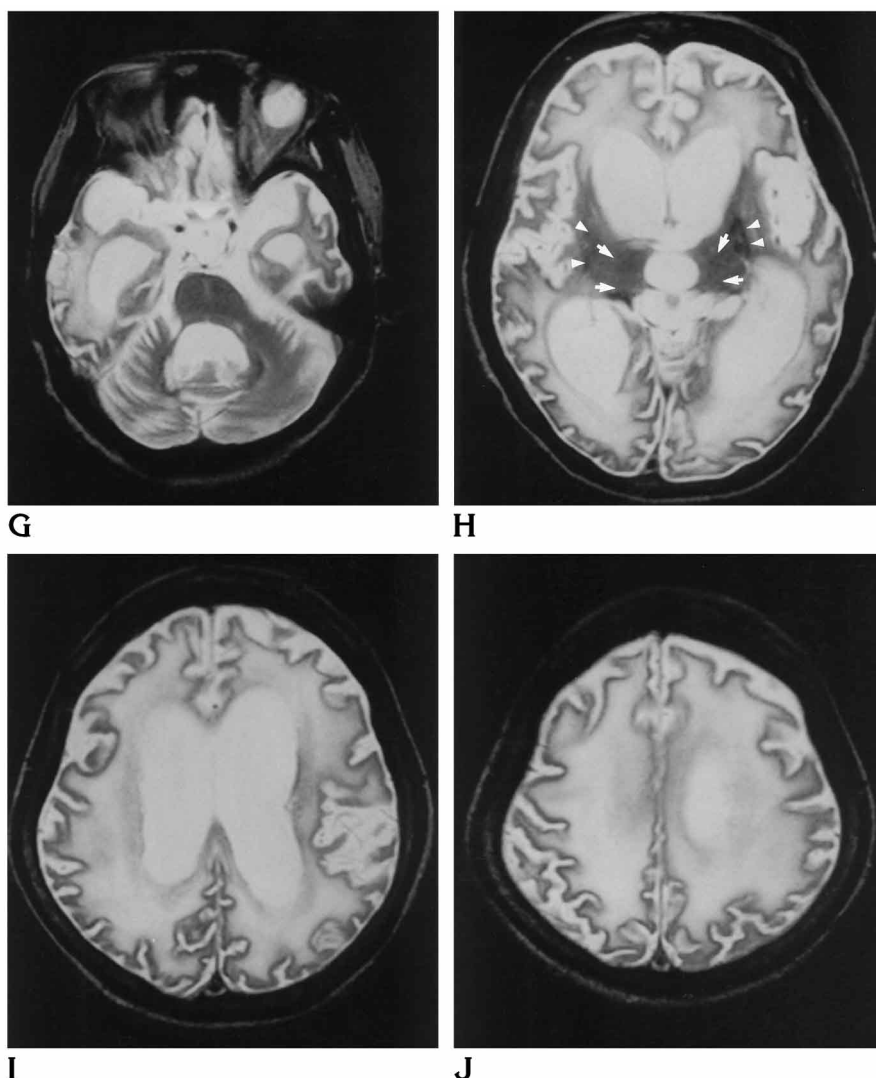
Fig 1, *cont'd.*

G–J, Axial MR images obtained 1 year after initial MR examination.

G, T2-weighted MR image (3000/90) shows marked cerebellar cortical atrophy and normal signal intensity of the brain stem.

H, At the level of basal ganglia, note marked atrophy of the cerebral cortex, basal ganglia (*arrowheads*), and thalami (*arrows*), with dilatation of the lateral and third ventricles. The cerebral white matter is of high signal intensity.

I and J, Cortical atrophy and demyelination are present at the levels of the body of the lateral ventricles and centrum semi-ovale.



Previous MR imaging reports of CJD have focused on abnormalities of the basal ganglia and cerebral cortex (9–11), yet involvement of focal occipital gray and white matter also have been reported (3–6). In 1992, Falcone et al (3) reported focal symmetrical occipital cortical involvement in CJD. Involvement of the cerebral white matter has been reported by a few authors. Kruger et al (4) reported a panencephalopathic type of CJD with extensive involvement of white matter showing periventricular signal change on T2-weighted MR images. Kawata et al (5) reported a case of CJD in which CT scans showed decreased attenuation in the cerebral white matter. Yamamoto and Morimatsu (6) commented that T2-weighted MR images may show high signal change in the basal ganglia in the early stage and diffuse hyperin-

tensity of the white matter in the terminal stage of CJD. The prevalence of hyperintense abnormalities in the basal ganglia on T2-weighted images in patients with proved CJD has not been studied and remains unknown. In our patient, the initial MR abnormalities were restricted to the basal ganglia (Fig 1B–E). Follow-up MR studies revealed significant and equal progression of the disease in the cerebral gray and white matter (Fig 1G–J). The diffuse T2 high signal in the white matter in our case was most likely the result of cortical neuronal death (Fig 1F), a phenomenon of diffuse wallerian degeneration.

Diffuse demyelination of cerebral white matter associated with cortical atrophy can also be seen in a variety of diseases such as Binswanger microangiopathy, human immuno-

deficiency virus encephalitis, the adult form of Pelizaeus-Merzbacher disease, and other neurodegenerative disorders (12–15). A history of progressive dementia with myoclonic seizures developing over a relatively short period in a middle-aged person may help in the differentiation. Open brain biopsy, however, is still mandatory for final confirmation of the diagnosis.

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