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Lateralized clinical and diffusion-weighted MRI abnormalities in a probable case of sporadic Creutzfeldt-Jakob disease.

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Summary

The usual clinical profile of sporadic Creutzfeldt-Jakob disease (sCJD) is subacute dementia, motor dysfunction and myoclonus. Occasionally, some patients present atypical clinical features. We report a case of probable sCJD in a 73-year-old man with a rapidly progressive lateralized neurologic dysfunction of the left hemisphere. In a few weeks the clinical picture deteriorated dramatically to akinetic mutism and myoclonus. The 14-3-3 protein was positive in the cerebrospinal fluid. Diffusion-weighted (DWI) magnetic resonance imaging (MRI) revealed increased signal in the left cortical ribbon and deep gray matter corresponding to the clinical lateralization. He died 9 weeks after onset, autopsy was not performed. This case illustrates the correlation between the lateralized clinical and DWI MRI abnormalities in sCJD.

Keywords: Creutzfeldt-Jakob disease; Magnetic resonance imaging; Focal neurologic deficit

Introduction

Sporadic Creutzfeldt-Jakob disease (sCJD) is a rapidly progressive fatal prion disorder with typical clinical findings of dementia, motor dysfunction and myoclonus. Different clinical variants have been described. We report a case of probable sCJD presenting, without dementia at onset but, with an unilateral onset and a remarkable neuroimaging features illustrating the correlation between the lateralized and focal clinical and diffusion-weighted (DWI) magnetic resonance imaging (MRI) abnormalities.

Case report

A 73-year-old man without significant past medical history experienced a progressive lateralized neurologic dysfunction of the left hemisphere. After 6 weeks, neurological examination, at admission, revealed right hemiparesis, severe aphasia, mild dysarthria and very mild cerebellar ataxia. Computed tomography was normal. EEG showed only a diffuse theta and delta slowing predominant on the left side without perodic sharp wave complexes (PSWC). Cerebral MRI was performed 7 weeks after the onset of symptoms, which revealed increased signal in the cortical ribbon, putamen and caudate nucleus of the left hemisphere on DWI sequences (Fig. 1) corresponding to the lateralized clinical symptoms. T2-Weighted (T2W) and fluid attenuated inversion recovery (FLAIR) images showed moderately increased signal of the deep, left and right, gray matter (Fig. 2). Cerebrospinal fluid (CSF) examination was normal but the 14-3-3 protein was positive. The patient rapidly worsened with severe aphasia, right brachial paresis and myoclonus. Finally he developed an akinetic mutism and intractable myoclonus and died 9 weeks after clinical onset. Neither genetic sequencing of the prion protein gene was performed

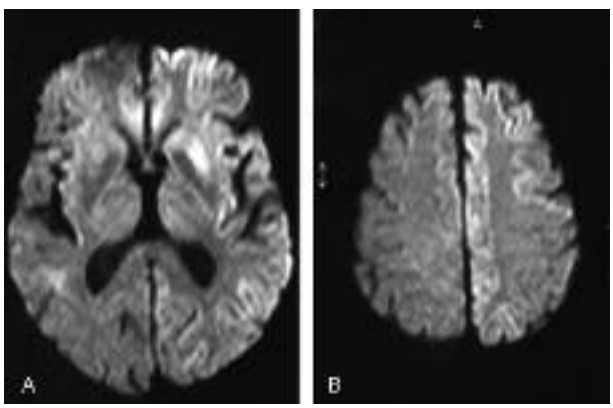

Fig. 1. — Axial DWI MRI (A-B) shows hyperintense signals in the left caudate nucleus and putamen as well as in the left cerebral cortex.

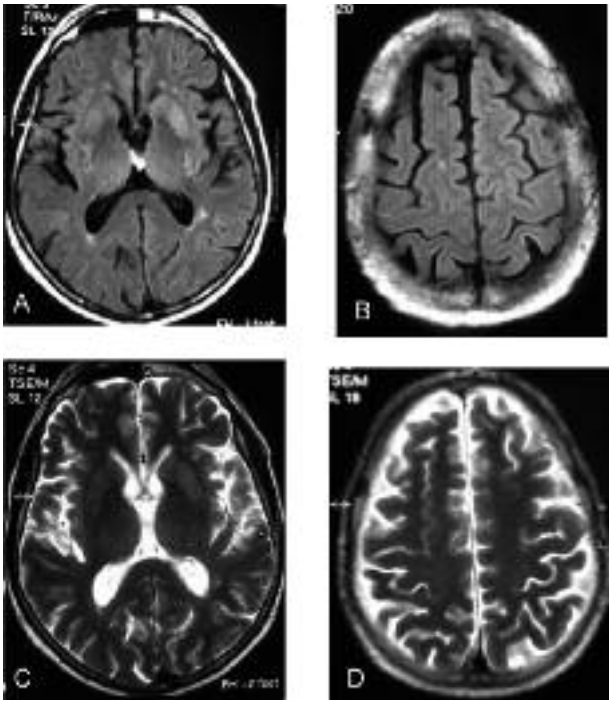

Fig. 2. — Axial FLAIR (A-B) and T2W MRI (C-D) images show bilateral subtle increase in signal intensity in the basal ganglia.

nor autopsy, because of the patient's family refusal. According to the World Health Organization (WHO, 1998) the final diagnosis in this case was thus "probable" sCJD.

Discussion

Classically, a progressive dementia lasting less than two years with at least two of four clinical features among which myoclonus, visual or cerebellar signs, pyramidal or extrapyramidal signs and akinetic mutism associated with an EEG showing generalized PSWC and/or a positive 14-3-3 protein test in CSF, provide the diagnosis of probable sCJD (WHO, 1998). CJD may sometimes, present with lateralized or focal cortical syndromes before the onset of more diffuse dementia or myoclonus with colocalizing EEG and MRI findings. Blasco Olcina et al. (2001) described an autopsy-proven sCJD with unilateral clinical onset and mesencephalic and focal cortical hyperintensity on FLAIR MRI. Cambier et al. (2003) reported a case series of 8 patients with sCJD who had a history of rapidly progressive lateralized or focal neurologic dysfunction, EEG, and corresponding abnormalities on FLAIR MRI.

Actually, MRI is not a criterion for the diagnosis of sCJD, although typical changes have been described. Traditionally, the recognised features of sCJD on MRI are bilateral symmetric markedly hyperintense caudate nuclei and putamina (Tschampa et al. 2005). Recent publications have additionally highlighted the importance of cortical ribbon hyperintensity (Shiga et al. 2004; Meissner et al. 2009). Shiga et al. (2004) described the MRIs of 36 patients with CJD (a mixture of definite and probable cases); 41,7% showed only cortical lesions, 45,8% showed lesions both in cortex and basal ganglia, and 12,5% lesions only in basal ganglia. The sensitivity of DWI was 92,3%, significantly more than T2W (41%) or FLAIR (50%). In 17 patients who did not show PSWCs on the first EEG, abnormal DWI findings were still clearly detected. DWI can be the only positive investigation in human prion disease (Shiga et al. 2004). It is the most sensitive test for the early clinical diagnosis of CJD as changes have been noted as early as 3 weeks after the onset of symptoms (Shiga et al. 2004). Considering the high sensitivity of brain MRI in the diagnosis of sCJD, Zerr et al. (2009) propose that the clinical diagnostic criteria should be amended to include MRI signal abnormalities.

Conclusion

The atypical presentation of focal or lateralized clinical findings may initially lead to an incorrect diagnosis. The correlation between lateralized clinical symptoms and DWI MRI abnormalities is helpful for the non-invasive diagnosis of sCJD, especially for the identification of atypical sporadic disease forms.

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