



Cortical Diffusion Restriction as the Single Abnormality on MRI in Creutzfeldt-Jakob disease

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Keywords

Creutzfeldt-Jakob disease, Sporadic CJD, Magnetic resonance imaging, Diffusion MRI.

Introduction

Creutzfeldt-Jakob disease (CJD) is a rare neurodegenerative disease characterized by rapidly progressive dementia, ataxia and myoclonus. MRI is an important tool in discriminating CJD from other dementias. Among mimics of CJD are rapidly evolving Alzheimer's disease, vascular dementia, frontotemporal dementia and paraneoplastic or immune-mediated limbic encephalitis. A CJD diagnosis can be missed and strong clinical suspicion is not always confirmed on autopsy [1]. Typical imaging findings are white matter abnormalities in the thalamus, the striatum and progressive atrophy [2,3]. We present two cases of CJD, both with atypical onset, in which the single abnormality on MRI consisted of a diffusion restriction (diffusion-weighted imaging) of part of the neocortex.

Case 1

A 62-year-old man presented with unsteadiness of gait, tingling in the feet and progressive cognitive impairment. On neurological examination the patient was dysarthric, smooth eye pursuit was impaired, there were sensory disturbances in the feet, extensor plantar responses and gait ataxia. A limbic encephalitis with polyneuropathy was suspected. Standard MRI sequences – performed by a 1,5 T Siemens scanner (axial T2 TSE, T2 BLADE, TIRM DF, axial T1 MPRAGE) – showed no abnormalities. Serological testing for paraneoplastic antibodies, including anti-Hu, proved negative. Repeated imaging one month later showed DWI hyperintensity of the cortex in the temporal, parietal and occipital region without pathology of the white matter (Figure 1). Background pattern on EEG was slow but without triphasic complexes. Cerebrospinal fluid showed an elevated tau level of 2400 ng/l and slight 14-3-3 protein positivity. The patient developed myoclonus and a diagnosis of CJD was made shortly before death. At autopsy sporadic CJD was confirmed.

Case 2

A 72-year-old man presented with apathy, aphasia and unilateral

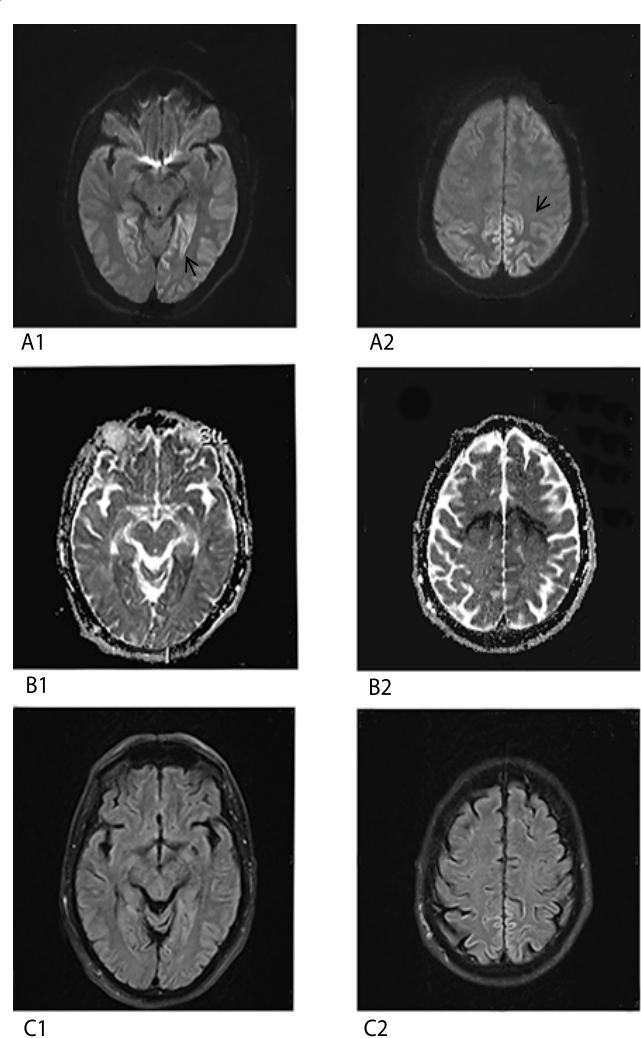


Figure 1: MRI in axial plane; DWI (A1,2): hyperintensity of temporal and occipital cortex (arrows). ADC map (B1,2): hypointensity of cortex. TIRM DF (C1,2): mild hyperintensity of cortex.

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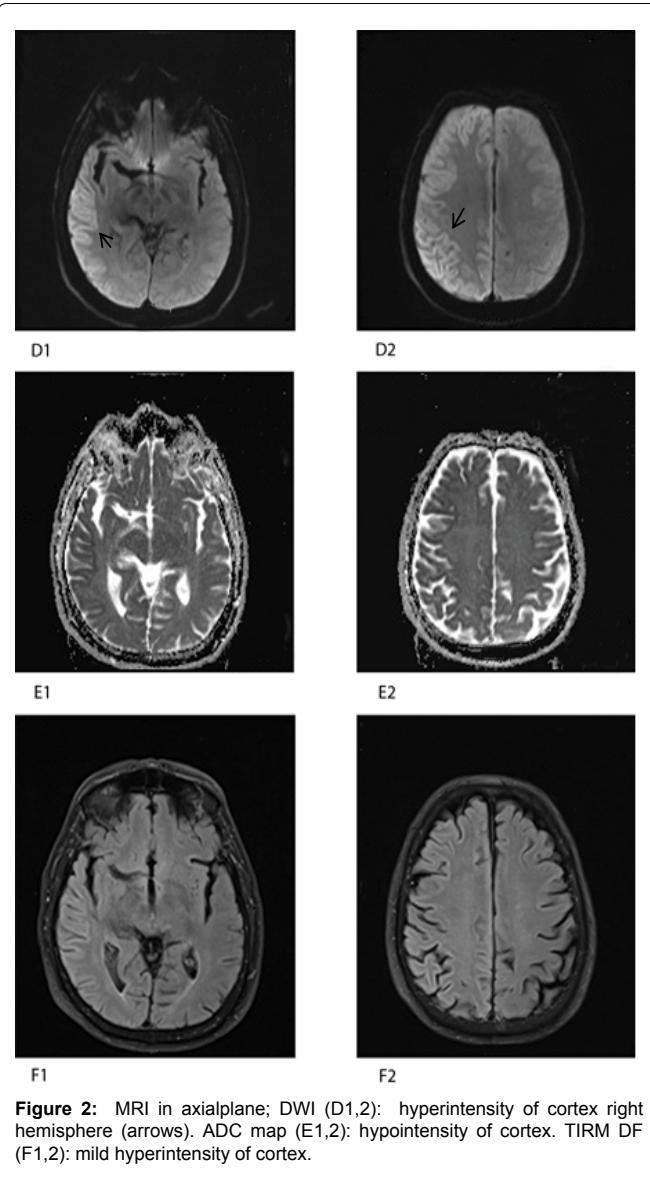


Figure 2: MRI in axial plane; DWI (D1,2): hyperintensity of cortex right hemisphere (arrows). ADC map (E1,2): hypointensity of cortex. TIRM DF (F1,2): mild hyperintensity of cortex.

weakness. There was a history of hypertension and an initial diagnosis of stroke was made. Standard MRI T2 sequencing showed a small, subcortical white matter lesion. The patient developed progressive ataxia and cognitive symptoms which were incompatible with the initial diagnosis. The EEG demonstrated a slow wave pattern with generalized triphasic complexes. Cerebrospinal fluid showed an elevated tau level of 9400 ng/l and 14-3-3 protein positivity. DWI cortical hyperintensity of the right temporal and occipital lobe was seen on a second MRI (Figure 2). The patient was diagnosed with probable CJD and died soon afterwards. Autopsy was not allowed.

Discussion

An early diagnosis of probable CJD can be difficult if signs are mild or atypical. Early differentiation from other causes of dementia is vital for determining prognosis and preventing unnecessary investigations. In case number one an anti-Hu paraneoplastic syndrome was considered more likely than CJD because of a new sensory neuropathy and cerebellar symptoms in a heavy smoker. The second patient was diagnosed with cerebrovascular disease at first. MRI led to a diagnosis of probable CJD. Diffusion restriction of the cortex was the single abnormality in 24% of cases in a study focussing on interrater characteristics [4]. DWI is found to have better sensitivity and specificity compared to other T2 weighted sequences in most studies [5]. New MRI criteria for CJD reflect the higher sensitivity of DWI compared to FLAIR/TIRM DF [6]. Since many standard MRI scanning protocols do not include DWI, cortical diffusion pathology can easily be missed so given the right index of suspicion the sequence should be included.

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