



Creutzfeldt-Jakob Disease in the Light of Diffusion Magnetic Resonance Imaging Findings

Difüzyon Manyetik Rezonans Görüntüleme Bulguları Işığında Creutzfeldt-Jakob Hastalığı

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Case

Fifty-seven year old male patient came to us with forgetfulness, sleepiness, hand tremor, sluggish movements and withdrawn mood. The patient's place, time and people orientation was disrupted in his neurological examination. There was postural tremor in the bilateral upper extremities. There was no pathological reflex. Routine biochemistry, hemogram, sedimentation, vitamin B12, thyroid function tests, cranial magnetic resonance imaging (MRI) and electroencephalography (EEG) were assessed as normal. He scored 13/30 points in Mini-mental state evaluation. While the second cranial MRI at the 45th day was normal, the diffusion restrictions in the frontal pole, anterior interhemispheric fissure and bilateral insular cortex seen in the diffusion-weighted imaging (DWI) attracted attention (Figures 1, 2). There was a widespread sluggishness in the EEG. Infection, autoimmune and paraneoplastic markers in the peripheral blood and cerebrospinal fluid (CSF) to investigate sub-acute onset dementia etiology were all negative. In terms of CSF, there were no cells, the biochemistry tests were normal, 14-3-3 protein was positive, tau protein and neuron-specific enolase values were normal. Neuropsychometric

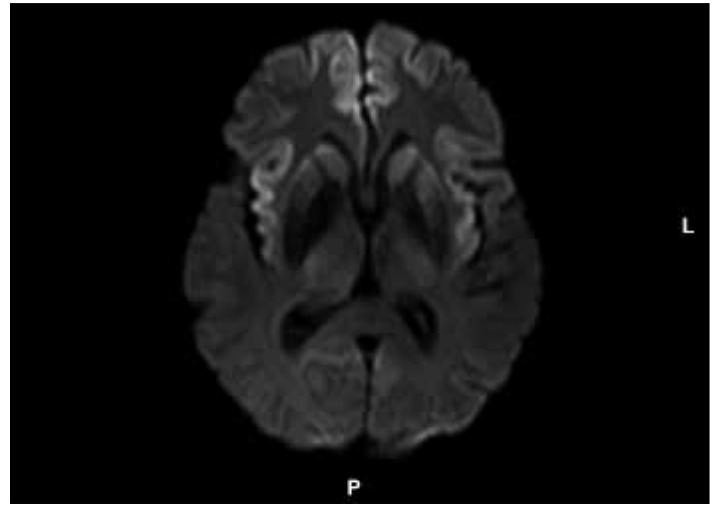


Figure 1. Diffusion-weighted image of ribbon sign in bilateral frontal and insular cortices

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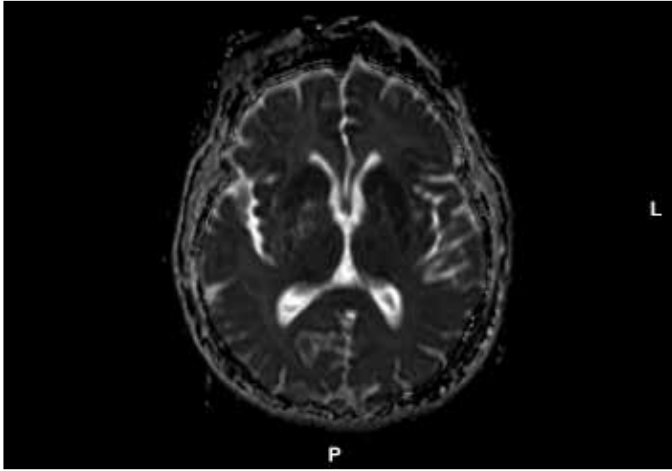


Figure 2. Apparent diffusion coefficient (ADC) image of ribbon sign in bilateral frontal and insular cortices

evaluation was compatible with frontal dysfunction that is characterized by global cognitive impairment. The patient was followed with Creutzfeldt-Jacob disease (CJD) diagnosis.

Imaging plays an important role in the diagnosis of dementia and in ruling out other conditions during the differential diagnosis. It was shown that the increased signal intensity seen in DWI has 92% sensitivity (1) for sporadic CJD. Increased signal strength in bilateral caudate and putamen in conventional diffusion-

weighted MRI is seen in 80% of the patients (2). Recent studies also emphasized the importance of the cortical ribbon sign in the diagnosis (1). For the diagnosis of sporadic CJD, DWI must satisfy one of the criteria below (3):

1- Abnormal unilateral or bilateral signal intensity in striatum and ribbon sign in one gyrus of the cerebral cortex.

2- Ribbon sign in more than 3 cortical gyri (in the absence of lesions in T1-weighted slices and white matter).

For the patients with predominant signal changes, these findings emerge within 10 weeks after the symptoms start and these cases were often associated with neuropsychiatric symptoms. Alien hand syndrome, dystonia, Parkinsonism and ataxia may also be seen (4). In our patient, the cortical ribbon sign seen in the frontal cortex facilitated the diagnosis of sporadic CJD.

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