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Isolated Cortical Involvement on MR imaging in Sporadic Creutzfeldt-Jakob disease: a case report

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ABSTRACT

Creutzfeldt-Jakob disease (CJD) is a rare dementing disease and is thought to caused by a prion. It is characterized by rapidly progressive dementia, ataxia, myoclonus, akinetic mutism and eventual death. The detection of 14-3-3 protein in the cerebrospinal fluid (CSF) may support the diagnosis of the CJD. Periodic synchronized sharp wave complexes are usually seen in electroencephalogram (EEG) during middle or late stages of disease. Diffusion-weighted imaging (DWI) is the most sensitive magnetic rezonans sequence technique in the diagnosis of CJD. Brain biopsy or autopsy is required for a definitive diagnosis of CJD.

We present 60-year-old man diagnosed as sporadic CJD with isolated cortical involvement. The patient had dementia and myoclonus. The 14-3-3 protein was positive in the CSF. Bilateral asymetric cerebral cortical abnormalities with high signal intensities were seen well on the DWI and to a lesser degree also seen on fluid-attenuated inversion recovery (FLAIR) images. No abnormal signal were seen in the basal ganglia and thalamus. Through this case report we want to emphasize the importance of DWI as a conjunct to the conventional MR sequences in cases in which the CJD is the prediagnosis in order to detect the disease in its earlier stages.

Key Words: Magnetic Rezonans Imaging, Creutzfeldt-Jakob disease, Isolated Cortical Involvement, Diffusion-Weighted Imaging

Sporadik Creutzfeldt-Jakob Hastalığında İzole Kortikal Tutulumun Manyetik Rezonans Görüntüleme Bulguları

ÖZET

Creutzfeldt-Jakob hastalığı (CJH) nadir bir demansiyel hastalıktır. Priona bağlı olduğu düşünülmektedir. Hızlı ilerleyen demans, ataksi, miyoklonus, akinetik mutizm ve sonrasında ölüm ile karakterizedir. Beyin omurilik sıvısı (BOS)' nda 14-3-3 proteinin tespiti CJH tanısına destek olabilir. Hastalığın orta veya geç evrelerinde elektroensefalografi (EEG)'de genellikle periyodik senkronize keskin dalga kompleksleri görülür. Difüzyon ağırlıklı görüntüleme (DAG) CJH tanısında en duyarlı manyetik rezonans görüntüleme (MRG) tekniğidir. Kesin tanı için beyin biyopsisi veya otopsi gereklidir. Biz bu yazıda olası sporadik CJH tanısı konan ve izole kortikal tutulumu olan 60 yaşındaki erkek olguyu sunuyoruz. Hastamızda demans bulguları ve myoklonus mevcuttu. BOS' da 14-3-3 protein pozitifti. Kranial MRG' de serebral kortikal bölgelerde DAG ve fluid-attenuated inversion recovery (FLAIR) görüntülerde bilateral asimetrik yüksek sinyal intensite izlendi. Bazal ganglionlar ve talamuslarda patolojik sinyal saptanmadı. CJH ön tanısı olan olgularda özellikle hastalığın erken dönemde saptamasında konvansiyonel MRG sekanslarına DAG eklenmelidir.

Anahtar kelimeler: Manyetik rezonans görüntüleme, Creutzfeldt-Jakob hastalığı, İzole kortikal tutulum, Difüzyon ağırlıklı görüntüleme

INTRODUCTION

Creutzfeldt-Jakob disease (CJD) is a rare dementing disease that often affects the younger age population than Alzheimer disease (dementia Alzheimer type, or DAT) which is usually seen in the seventh decade of life (1). CJD is caused by a small proteinaceous infectious agent devoid of DNA and RNA, called prion (1,2). The disease produced by the conversion of the prion protein molecule PrPC to PrPSC (scarpie particles) (1) and the accumulation of the pathologic scarpie particles (PrPSC) in the human brain (3,4). In the classic type, the disease is characterized by rapidly progressive dementia, ataxia, abnormal muscle tone, and myoclonus. After a few months the akinetic mutism develops and the

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patients typically died within a year (3). CJD has four types: sporadic, familial, iatrogenic and variant forms. Approximately 85% of prion-related diseases are sporadic and have an unknown route or source of infection (3).

Case report

60 year old man presented to the neurology department of our hospital with the symptoms of bizarre talking, social withdrawal, weakness and involuntary movements in his right arm. As learned from his relatives, all these symptoms had started 2 months ago. One month ago, with the complaints and the symptoms of the weakness in his right arm and impediment in speech, the patient was hospitalized; and several possible diagnosis such as encephalitis, cerebrovasculer disease or CJD assumed. There was no high temperature nor stiff neck. Myoclonus and the signs of dementia detected clinically. Biochemical and hemogram profile, tiroid hormone levels, biomarkers of vasculitis were all within normal limits. The cerebrospinal fluid (CSF) biochemical profile was as follows: protein: 54 mg/dl, glukoz: 78 mg/dl, clor 118 mmol/L, LDH 53 U/L, Na 145 mmol/L, K 2,9 mmol/L and no bacteria-nor other cell types detected. The CSF culture yielded in no viral or bacterial proliferation. However the CSF was positive for the 14-3-3 protein. An epileptiform focus in frontotemporal area leading to high degree of secondary generalization in addition to low level but

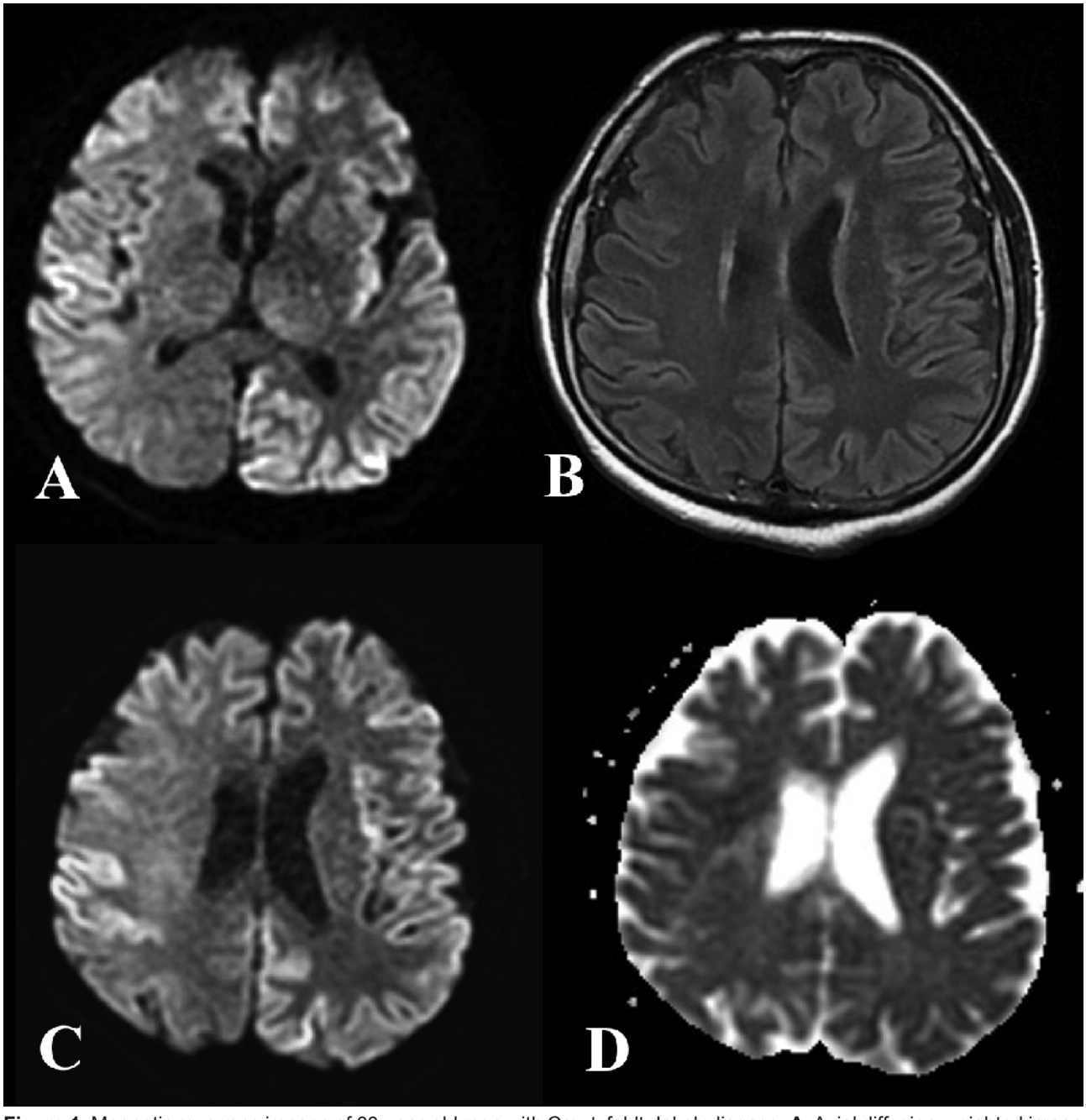

Figure 1. Magnetic rezonans images of 60-year-old man with Creutzfeldt-Jakob disease. **A.** Axial diffusion-weighted image obtained at level of basal ganglia shows abnormal high signal intensities in bilateral cerebral cortical regions.Axial fluid-attenuated inversion recovery image (**B**) and diffusion-weighted image, **C** shows abnormal high signal intensities in bilateral cerebral cortical regions. Apparent diffusion coefficient map, **D** shows low signal in the abnormal high signal intensity regions observed on the diffusion-weighted image.

widespread disturbance in cerebral bioelectrical activity detected in electroencephalogram (EEG).

Computerized brain tomography (CBT) findings was normal. Cranial magnetic rezonans imaging (MRI) examinations on the T1- weighted images (T1WI) and T2-weighted images (T2WI) were normal. However, especially more prominent on the diffusion-weighted imaging (DWI) and also to some degree on fluid-attenuated inversion recovery (FLAIR) images, abnormal high signal intensities unmatched to arterial vascular distrubition in the cortical frontal and parietal regions were seen in an assymmetric fashion and bilaterally. The apparent diffusion coefficient (ADC) values were low in the abnormal high signal intensity regions observed on the DWI (Figs. 1A-D). No abnormal signal were seen in the basal ganglia and thalamus. No pathological enhancement was detected following contrast media injection. From all these findings the diagnosis of sporadic CJD assumed with high probability. Our patient died one month after admission.

DISCUSSION

CBT studies may be normal (%80) or may show rapidly progressive atrophic changes in the brain (1). Rapidly progressive brain atrophy and high signal intensities in the cerebral cortex and the basal ganglia are well known MRI findings (3). Similar MR findings can also be seen in the thalamus and cerebellum (3,4). DWI is the most sensitive MRI technique in the diagnosis of CJD (5). Even though the DWI is important in the earlier diagnosis of CJD, its value may be somewhat limited in the more advanced stages of the disease (3).

Meissner et al. made a study of 55 patients with two major lesion patterns were identified by DWI: cortex and basal ganglia involvement (two thirds) and isolated cortex involvement (one third). In the isolated cortex involvement, it was usually the frontal and parietal lobes that effected (%78) (4). In our patient DWI and FLAIR images showed abnormal bilateral asymmetric high signal intensities in cortical regions of the frontal and parietal lobes (being more prominent on the DWI).

Differentiation from venous hypertensive encephalopathy, hypoxia, epilepsy and encephalitis may be necessary, because they also present with abnormal cortical high signal intensities in DWI (6,7,8,9).

The detection of 14-3-3 protein in the CSF may support the CJD diagnosis, but this protein may be seen in other central nervous system disorders such as viral encephalitis, Hashimoto encephalitis, amyotrophic lateral sclerosis, and other types of dementia and it is not pathognomonic of CJD (3,4,10,11). In our case, the CSF was positive for the 14-3-3 protein.

EEG does not give any spesific clue regarding to the CJD but only nonspesific manifestations such as normal or diffuse slowing, and frontal rhythmic delta activity may be seen. Periodic biphasic or triphasic, synchronized sharp wave complexes occuring during middle or late stages of disease are typical and found 90% of the patients (10).

Brain biopsy or autopsy is required for the definitive diagnosis of CJD (12).

In conclusion, clinical, laboratory, EEG and MRI findings largely support the diagnosis of CJD. Sporadic CJD can also present with isolated cortical involvement and in cases in which the CJD is the prediagnosis, beside conventional sequences DWI should be added to the routine MRI examination in order to detect the disease in its earlier stages and so the unnecessary biopsy should be avoided.

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