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Images

Diffusion-weighted imaging MRI in Creutzfeldt-Jakob disease

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1. Case description

1.1. Patient one

A 67-year-old female with hypertension, autoimmune hypothyroidism and history of epilepsy, presented with a two-month history of progressive cognitive compromission (deficit of short time memory, hallucinations, confusion).

Brain MRI showed in diffusion-weighted-imaging (DWI) and in fluid-attenuated-inversion-recovery (FLAIR) sequences an alterated signal in parieto-fronto-temporal cortices (Fig. 1).

1.2. Patient two

A 55-year-old male with hypertension, rheumatoid arthritis, presented with a seven-day history of progressive gait imbalance until use of wheelchair.

Brain MRI showed in diffusion-weighted-imaging (DWI) and in fluid-attenuated-inversion-recovery (FLAIR) sequences an alterated signal in occipito-parieto-temporal cortices (Fig. 2).

1.3. Patient three

A 69-year-old female with dyslipidemia, presented with a three-month history of progressive behavioral disorder, gait imbalance until use of wheelchair, paresis of left limbs.

Brain MRI showed in diffusion-weighted-imaging (DWI) and in fluid-attenuated-inversion-recovery (FLAIR) sequences an alterated signal in right parieto-frontal cortices and in putamen (Fig. 3).

1.4. Patient four

A 71-year-old male, smoker, with arrhythmic-hypertensive heart disease, presented with a ten-day history of dizziness with progressive balance instability, cerebellar syndrome, with cognitive impairment.

Brain MRI showed in diffusion-weighted-imaging (DWI) and in fluid-attenuated-inversion-recovery (FLAIR) sequences an alterated signal in left occipito-parietal cortices, bilateral frontal cortex and in left striatum (Fig. 4).

The four patients underwent also to brain MRI, EEG, cerebral spinal fluid (CSF) examination.

2. What is your diagnosis?

a. Wernicke's encephalopathy
b. Viral encephalitis
c. Creutzfeldt-Jakob disease
d. Hypoxic ischemic encephalopathy

3. Diagnosis

Creutzfeldt-Jakob disease.

4. Discussion

We considered 4 cases of suspicious Creutzfeldt-Jakob disease (C.J.) The onset symptoms have often not been characteristic, becoming progressively more specific.

C.J. is prion encephalopathy, characterized by progressive dementia with cognitive, behavioral and/or motor dysfunction; it is a rare disease, usually fatal within 12 months after onset; there are several forms of C.J: the most common form is sporadic (SCJD); other forms are familial or genetic. Diagnosis of the disease is based on the clinical, usual EEG pattern, detection of protein 14.3.3 in CSF, and MRI; the definitive diagnosis requires brain autopsy.

EEG of four patients showed a typical pattern with periodic or pseudoperiodic sharp-waves complexes (PSWCs). CSF examination documented the presence of 14.3.3 protein in all patients.

In the first phase of pathology, our patients had a cerebral cortex involvement, and only in one there was an involvement of basal ganglia.

Brain MRI is the gold standard in C.J. diagnosis thanks to its greater sensitivity in T2-FLAIR and DWI than in "classic" long TR sequences. MRI typically showed an T2-weigted and FLAIR

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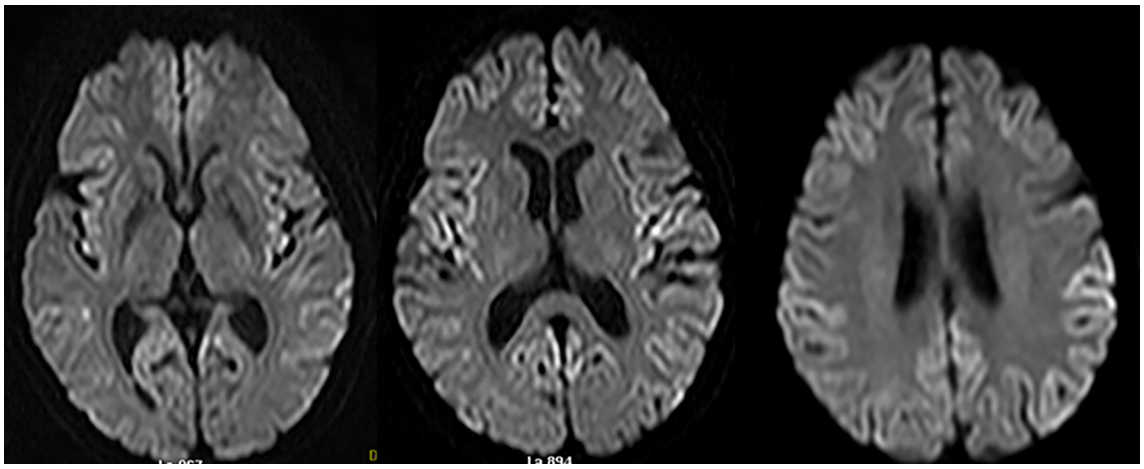
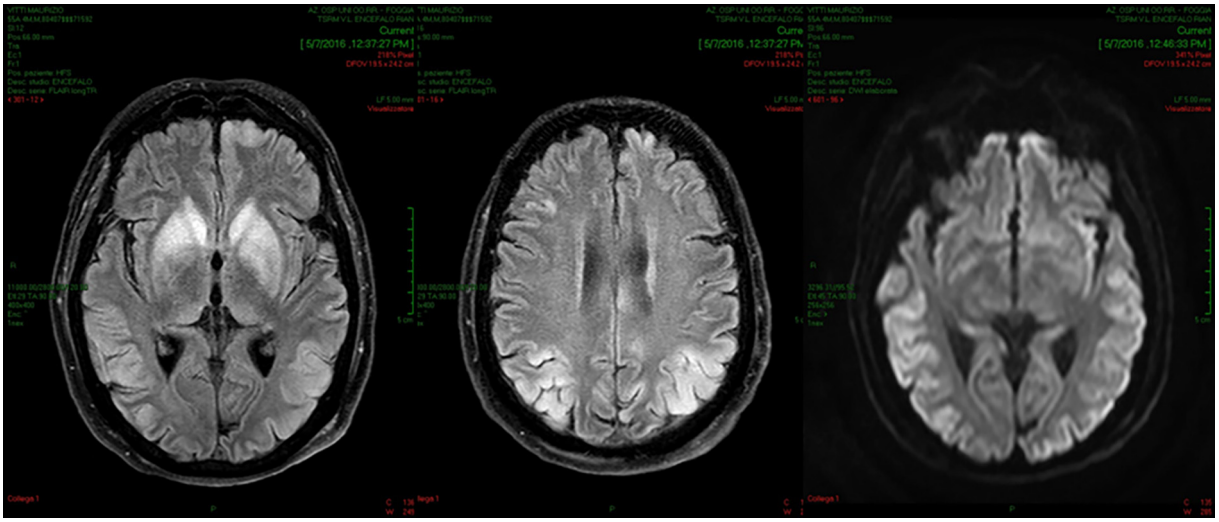


Fig. 1.



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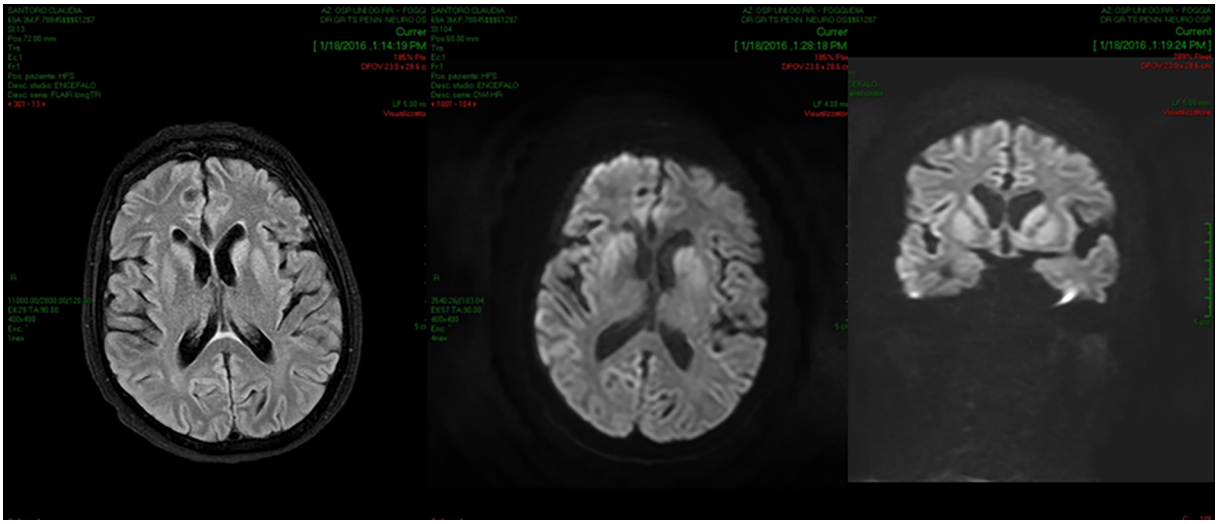

Fig. 3.

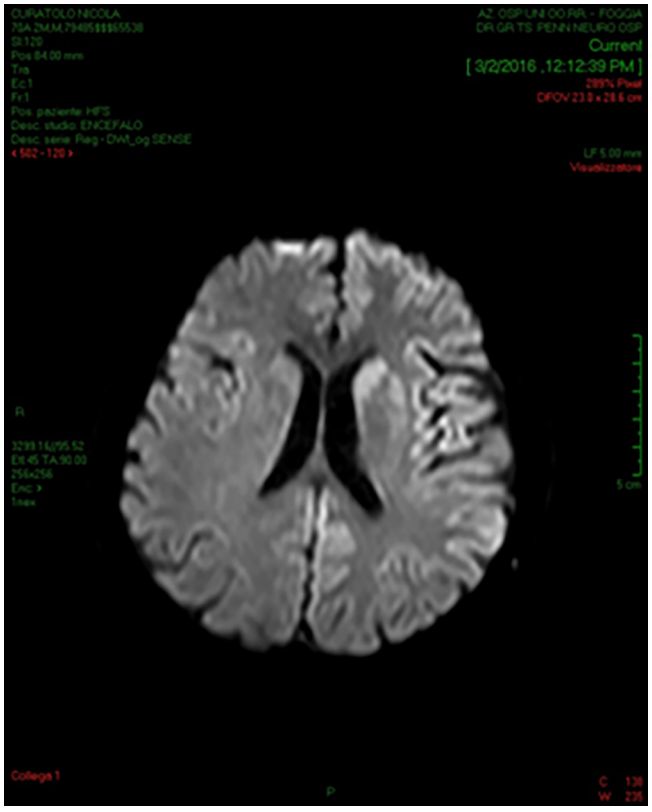

Fig. 4.

alterated signal in basal ganglia; the possible involvement of the cerebral cortex requires the use of DWI sequences. DWI with Apparent Diffusion Coefficient (ADC) mapping showed an increased sensitivity, because of better contrast in cortex study.

Conflicts of interest

We declare that there are no conflicts of interest. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.