CJD micking NCSE: can the ictal genesis of epileptiform abnormalities always be ruled out? EEG features and discussion of a case report.

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Introduction

Creutzfeldt–Jakob disease (CJD) is a prion-related neurodegenerative disorder. The interpretation of epileptic activity, particularly non-convulsive status epilepticus (NCSE), in CJD remains controversial. We report a case of CJD presenting with NCSE and discuss the importance of determining the ictal nature of electroencephalographic (EEG) abnormalities.

Materials and methods

We conducted a comprehensive PubMed search to identify CJD cases presenting with status epilepticus (SE). Search terms included combinations of “CJD,” “SE,” “NCSE,” “seizures,” “EEG,” and “prion.” Priority was given to cases where NCSE was an early feature of CJD.

Results

An 83-year-old woman without previous cognitive impairment or relevant medical history presented with a 2-week history of episodic confusion, bizarre behavior, progressive gait instability and clumsiness. On admission, confusion, disorientation, persevering behavior, fluctuating global aphasia, upward gaze limitation and extrapyramidal signs were observed. EEG showed 2-2.5 Hz periodic generalized sharp- and spike-waves, predominant in the right temporal region and briefly interrupted by sensory stimulation. Diazepam and levetiracetam administration abolished the periodic pattern without clinical improvement, fulfilling possibile NCSE criteria. Brain magnetic resonance demonstrated hyperintensity in the right caudate and putamen in DWI sequences and DWI and FLAIR hyperintensity in the right temporo-parietal cortical ribbon. Subsequent lumbar punctures showed altered neurodegenerative markers (Tau >2000 pg/ml) and RT-QuIC was positive, confirming probable CJD. The patient rapidly deteriorated and died two weeks later.

Discussion

The interpretation of EEG abnormalities in CJD remains controversial. While ~15% of sporadic-CJD cases have seizures in advanced stages, presentation with seizures is rare (~3%) and only few cases of NCSE have been documented. Typical CJD EEG patterns, like periodic sharp-wave complexes (PSWCs), periodic lateralized discharges (LPDs), stimulus-induced rhythmic or periodic ictal discharges (SIRPIDs) and generalized low voltage slowing, depend on the stage of the disease and may share overlapping features with NCSE. The early stages LPDs may transiently respond to antiseizure medications, suggesting possible ictal activity, but their frequent resolution with sleep or sensory stimulation supports their interpretation as non-ictal disease-related phenomena rather than seizure interruption. Our case met modified Salzburg criteria for possible NCSE with lack of clinical improvement. Moreover, EEG abnormalities often ceased with stimulation, complicating interpretation. Recent literature emphasizes the diagnostic ambiguity of such patterns in CJD and warns against overinterpreting isolated pharmacologic responses.

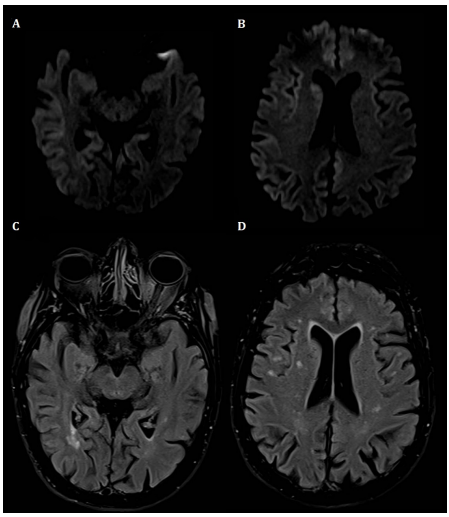

Figure 1. Modest diffusion restriction in the cortical strip in the right middle and superior temporal gyrus, right angular gyrus, right supramarginal gyrus, both cingulate gyri, head and body of the right caudate nucleus, and a small portion of the anterior putamen (A-B). Blurred FLAIR hyperintensity of the cortical ribbon in the temporo-parietal region without swelling (C-D).

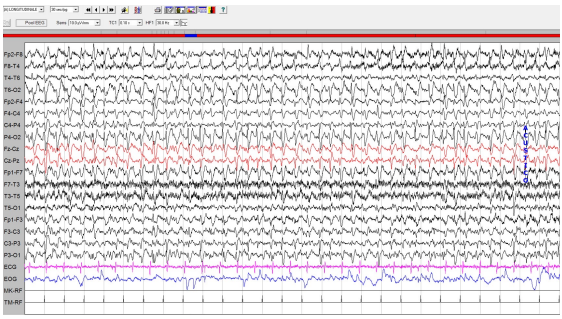

Figure 2. EEG showing right fronto-parieto-temporal spike-waves and sharp-waves at 2-2.5 Hz with spread to the contralateral homologous regions.

Conclusions

This case represents a rapidly-progressive CJD with early EEG features mimicking NCSE. Treating such EEG patterns remains debatable, but an antiseizure trial is advisable while awaiting diagnostic confirmation of the underlying unfavorable pathology.

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