CLINICAL IMAGES

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Periodic Discharges on Eeg in An Elderly Female with Vision Loss

ABSTRACT

Introduction: Rapidly progressive dementia includes a heterogeneous group of disorders. Electroencephalography (EEG) is one of the most important diagnostic tests, especially in the context of disorders such as Creutzfeldt–Jakob disease (CJD), autoimmune encephalitis, metabolic encephalopathy, and non-convulsive status epilepticus.

Case Report: A 78-year-old female with no known comorbidities presented with painless, progressive diminution of vision in both eyes over 4 months, followed by abnormal involuntary movements and rapidly progressive cognitive decline. She was referred to our centre after showing no improvement with empirical immunotherapy. On examination, she had altered sensorium, paucity of movements on the right side, and myoclonic jerks. EEG was performed and showed generalized theta slowing with stereotyped periodic sharp wave complexes (PSWCs). Magnetic resonance imaging (MRI) of the brain with diffusion-weighted imaging demonstrated diffuse asymmetric cortical restricted diffusion involving the bilateral cerebral cortices (left more than right), consistent with cortical ribboning. Cerebrospinal fluid (CSF) analysis was positive for the 14-3-3 protein. A diagnosis of sporadic CJD was made.

Discussion: The main EEG abnormality in sporadic CJD is the presence of periodic discharges (PSWCs). EEG abnormalities vary depending on stage of the disease. Periodic discharges that mimic CJD can occur in metabolic encephalopathy, autoimmune encephalitis, lithium toxicity, anoxic injury, and sometimes in nonconvulsive status epilepticus; however, these typically lack the highly stereotyped 1-Hz periodicity seen in sporadic CJD, particularly when correlated with the clinical progression.

Conclusion: EEG has significant clinical utility. It helps distinguish CJD from autoimmune encephalitis and nonconvulsive status epilepticus, in patients with rapidly progressive dementia when used in conjunction with MRI and CSF prion assays.

Keywords: Creutzfeldt–Jakob disease; cortical ribboning; EEG; Rapidly progressive dementia

INTRODUCTION

The term "rapidly progressive dementia" refers to a subgroup of dementias in which progression from the first symptom to the onset of dementia occurs over an interval of weeks to months, with the majority of patients progressing from independence to complete dependence within 1–2 years.[1,2] These account for approximately 3–4% of all dementia cases in clinical practice and therefore represent a rare subgroup.[1-3] As a group, these disorders are diagnostically challenging, as the differential diagnoses are wide-ranging and include vascular, infectious, toxic-metabolic, neurodegenerative (including prion disease), autoimmune, and neoplastic causes, among others.[3]

Electroencephalography (EEG) is one of the most important diagnostic tests, particularly in disorders such as Creutzfeldt–Jakob disease (CJD), autoimmune encephalitis, metabolic encephalopathy, and non-convulsive status epilepticus.[4] In CJD, especially when advanced cerebrospinal fluid biomarkers or real-time quaking-induced conversion (RT-QuIC) assays are not available, EEG can serve as a robust diagnostic aid.[5] We present the case of a 78-year-old female with vision loss and rapidly progressive cognitive decline in whom EEG demonstrated characteristic periodic discharges suggestive of CJD. We highlight this distinctive EEG finding.

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CASE HISTORY

A 78-year-old female with no known comorbidities presented with painless, progressive diminution of vision in both eyes over a period of 4 months. She subsequently developed abnormal involuntary jerky movements involving the limbs. This was followed by rapidly progressive cognitive decline characterized by confusion, reduced interaction, and impaired responsiveness.

She was initially evaluated at an outside hospital. Magnetic resonance imaging (MRI) of the brain with orbit study was reported as normal. Cerebrospinal fluid (CSF)

examination revealed mildly elevated protein (56.2 mg/dL), glucose of 96 mg/dL, and 2 cells/mm³. Extensive workup, including infectious and autoimmune evaluation, was negative. This included CSF studies for tuberculosis, bacterial cultures, cytology, and an autoimmune encephalitis panel. Serum vasculitic markers were also negative. With a suspicion of autoimmune encephalitis, she received immunotherapy consisting of intravenous methylprednisolone (1 g daily for 5 days) followed by intravenous immunoglobulin (IVIG) (2 g/kg administered over 5 days) empirically. However, there was no significant clinical improvement.

She was subsequently referred to our centre for further management. On examination, she was drowsy and intermittently arousable to verbal commands. She had bulbar dysfunction and was on Ryle's tube feeding. Motor examination revealed increased tone in all four limbs with brisk deep tendon reflexes and relative paucity of movements on the right side. Myoclonic jerks were also noted.

Electroencephalography (EEG) was performed and showed generalized theta slowing with stereotyped periodic sharp wave complexes at approximately 1 Hz, predominantly posteriorly distributed, with intermittent phases of electrical quiescence (Figure 1). These findings were suggestive of Creutzfeldt–Jakob disease (CJD). MRI brain diffusion-weighted imaging demonstrated diffuse asymmetric cortical restricted diffusion involving the bilateral cerebral cortices (left more than right), consistent with cortical ribboning (Figure 2). CSF analysis was positive for the 14-3-3 protein. Based on these findings, a diagnosis of sporadic CJD (Heidenhain variant) was made.

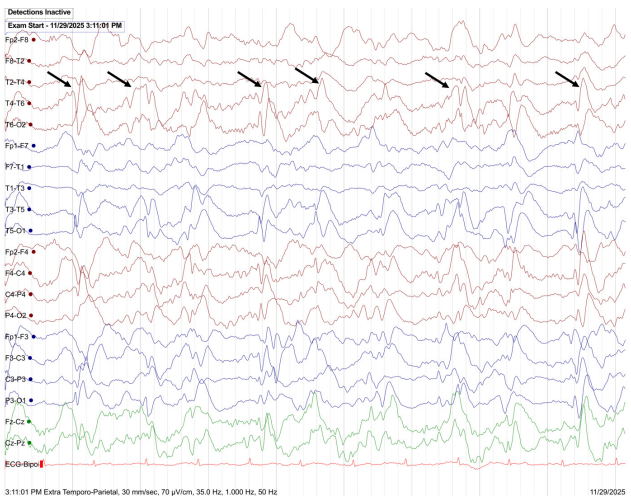

Figure 1: EEG recording showing generalised theta slowing with stereotyped periodic sharp wave complexes at approximately 1 Hz (black arrows), predominantly posteriorly distributed, with intermittent phases of electrical quiescence, findings suggestive of Creutzfeldt–Jakob disease (CJD).

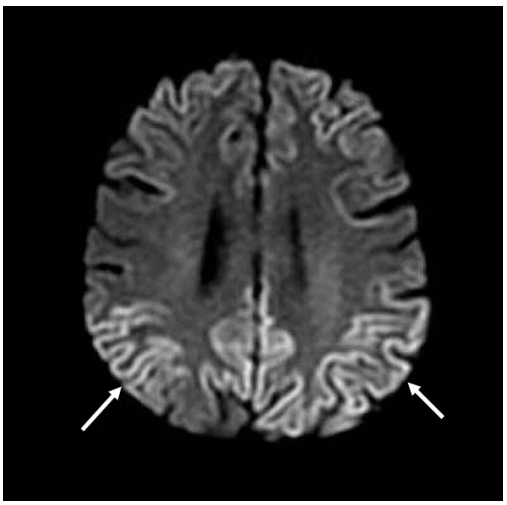

Figure 2: MRI Brain diffusion weighted imaging showed diffuse asymmetric cortical restricted diffusion involving bilateral cerebral cortices (left more than right) consistent with cortical ribboning (white arrows)

DISCUSSION

The main EEG abnormality in sporadic CJD is the presence of periodic sharp wave complexes (PSWCs), which are high-amplitude biphasic or triphasic sharp waves occurring at regular intervals, usually every 0.5–2 seconds (classically ~1 Hz).[5-7] These complexes are most noticeable in the frontocentral regions but usually demonstrate a diffuse cortical distribution. Morphologically, they consist of sharply contoured discharges lasting 100–300 ms, often superimposed on a diffusely slowed background. As cortical neuronal loss advances, the background rhythm is progressively replaced by theta and delta activity.[5]

EEG abnormalities evolve with the stage of the disease. In the early stages, the tracing frequently reveals only nonspecific diffuse slowing, occasionally accompanied by frontal intermittent rhythmic delta activity. PSWC typically appear a few weeks after the onset of symptoms as neuronal damage progresses.[7] Their presence correlates strongly with the akinetic-mutism phase and with cortical ribboning on diffusion-weighted MRI. Notably, PSWC are rarely observed at an early stage and may disappear when cortical electrical activity markedly decreases in the terminal stage.[5,8]

The diagnostic performance of PSWC is characteristic but not perfect. Reported sensitivity ranges from 44–66%, whereas specificity exceeds 85–90% when interpreted in the appropriate clinical context.[6,9] However, the absence of PSWC does not exclude CJD, particularly in certain molecular subtypes such as MV2 or VV2, in younger patients, and in variant or genetic prion diseases.

Periodic discharges that mimic those seen in CJD may occur in metabolic encephalopathy, autoimmune encephalitis, lithium toxicity, anoxic injury, and occasionally in non-convulsive status epilepticus. However, these conditions typically lack the highly stereotyped 1-Hz periodicity observed in sporadic CJD, particularly when correlated with the characteristic clinical progression.[7,9]

Pathophysiologically, PSWC are thought to represent oscillatory activity generated by dysfunctional thalamo-cortical circuits. Prion-mediated spongiform degeneration preferentially disrupts inhibitory interneurons, resulting in hypersynchronous cortical firing. The periodicity likely reflects rhythmic thalamic pacemaker activity imposed on a globally hyperexcitable cortex.[5,10] This model may explain the association between PSWC, myoclonus, and cortical diffusion restriction seen on MRI.

CONCLUSION

EEG has significant practical clinical utility in rapidly progressive dementia, as it helps distinguish CJD from conditions such as autoimmune encephalitis and non-convulsive status epilepticus. It can also guide anti-epileptic treatment in patients with prominent myoclonus and assist in prognostication, as the appearance of PSWC generally indicates advanced cortical involvement and is associated with a shorter survival interval.

Clinical Significance

Characteristic PSWC on EEG strongly support the diagnosis of sporadic CJD when interpreted in conjunction with diffusion-weighted MRI findings and CSF prion biomarkers.

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