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Case Report

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Sporadic Creutzfeldt-Jakob Disease Presenting as Nonconvulsive Status Epilepticus Case Report

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Keywords:

Creutzfeldt-Jakob disease; status epilepticus.

1. Abstract:

Creutzfeldt-Jakob disease (CJD) is the most common transmissible spongiform encephalopathy in human. It is characterized by triad of dementia, myoclonus and periodic sharp complex waves on EEG, with this easy to be recognized features, CJD diagnosis is challenging especially if present with atypical presentation. We report a patient with non- convulsive status epilepticus.

2. Introduction

Creutzfeldt-Jakob Disease (CJD) is the most common transmissible spongiform encephalopathy in human with average age of 62. The initial symptoms can be confusion, short-term memory loss, behavioral changes or neurological focal deficits such as ataxia. Subsequently, the disease evolves with rapidly progressive dementia and cortical myoclonus and finally akinetic mutism (triad of dementia, myoclonus and periodic sharp complex waves on EEG [1, 2]. It occurs as sporadic, genetic, and transmissible disease in humans. Sporadic (s) CJD accounting for approximately 85% of cases [3]. Researchers speculate that it results from a spontaneous neurodegenerative illness, and the hypothesis is that it results from either a somatic mutation in the gene or a random structural change in the PrP protein causing formation of PrPSc. Onset usually occurs in the 7th decade of life, and the median time to death is 5 months, with 90% of patients' dead by 1 year [4]. Nonconvulsive status epilepticus is characterized by continuous epileptiform patterns on electroencephalogram (EEG), alteration of consciousness including coma, and the absence of convulsive motor activity [5].

Few cases were reported with non- convulsive status epilepticus as presenting symptoms of sporadic CJD [6].

We report a patient with CJD who developed nonconvulsive SE. Continuous EEG monitoring was performed and revealed an EEG ictal pattern of PSWC

3. Case Report

65 years old Saudi male known case of diabetes mellitus came with history of 3 weeks' changes in his behaviors, unsteadiness, slurred speech and episodes of staring around. They noticed also intermittent brief rapid involuntary jerks involving one limb at a time, suggestive of myoclonic jerks, he developed intermittent abnormal twisted posturing of her right hand, suggestive of focal dystonia. He did not have any other previous or current drug or alcohol use, which was corroborated by his family. He had no significant family history. Patient is using Glucophage for his diabetes.

He is setting on bed uncomfortable, moving side to side, looking to objects around the bed with poor eye contact. Reduced attention span, impairment of short-term memory, behavioral abnormality, and language problems in the form of comprehension difficulty as well as irrelevant talking were the major features at the onset of her cognitive disturbance.

Conscious alert but nor oriented to time nor place.

Bouts of agitation were noticed during examination. Vital signs within normal limits

Patient speaking non fluently poor letters outcomes, poor comprehensive, impaired repetition, impaired writing, reading, and naming.

Other elements of higher mental functions were difficult to be done

due to poor patient cooperation.

Cranial nerves were intact. patient gait was normal. Normal power, tone, and reflexes.

Normal sensory to pinprick other modalities of sensation cannot be done. Normal coordination (Table 1).

MRI brain showed fluid attenuated inverted recovery (FLAIR) signal abnormality and diffusion restriction of bilateral striatum and scattered area at right tempo occipital region and bilateral cingulate cortices resembling ribbon pattern (Figure 1-3).

Urgent EEG showed repetitive theta activity on the left with gradual evolution consistent with seizure. Based on these results, he was treated as a case of status epilepticus with IV midazolam infusion and anti- epileptic drugs loading along with intubation and transferred to ICU. Lumber puncture done and result was normal for cells, protein and glucose. His seizure was difficult to control, so multiple EEG monitoring sessions were carried out. One of the multiple EEGs showed generalized periodic discharges per one second in form of sharp waves intermixed with slow activity 3-4 Hertz.

Later, EEG monitoring shows generalized periodic discharge GPDs 1-2/sec intermixed with rhythmic slow wave discharge with evolution with poor response to treatment. A diagnosis was provisionally established based on the positive result of 14-3-3 protein an RT-QuIC detection in CSF.

Patient was treated as status epilepticus reaching intubation and sedative agent with midazolam, we also started him on levetiracetam, valproic acid and phenobarbitone till his seizure subsided.

Tracheostomy was placed and situation explained to the family do not resuscitate form was signed. After 3 months' patient passed away.

Our patient had an atypical initial clinical presentation but with progressive changes in his radiological, EEG, and CSF findings becoming more typical and diagnostic for CJD. The progression of his imaging (MRI and EEG) and laboratory (CSF) abnormalities correlated well with his neurological deterioration. This emphasizes that uncommon etiologies must be considered in cases of unexplained and rapid neurological deterioration, especially with altered mentation and/or presence of refractory seizure activity. Close clinical monitoring is prudent and needs to be correlated with neuroimaging and cerebrospinal fluid analysis, which might aid in the early diagnosis of this lethal condition.

Table

Labs:	Result:
CBC	normal
Renal profile	normal
Liver function tests	normal
Thyroid functiontests	normal
Vitamin B12,folateand D	normal
homocysteine level	normal
Erythrocyte sediment rate andC reactive protein	normal
Paraneoplasticpanel	normal
Syphilis, HIV,TB,HSV	negative
CSF glucose	4.2
CSF protien	0.24
CSF WBC and RBC	0
CSF culture	Negative for bacterial fungal orparasitic growth.
CSF protein 14-3-3	positive
CSF Real-time quaking-inducedconversion (RT- QuIC)	positive.

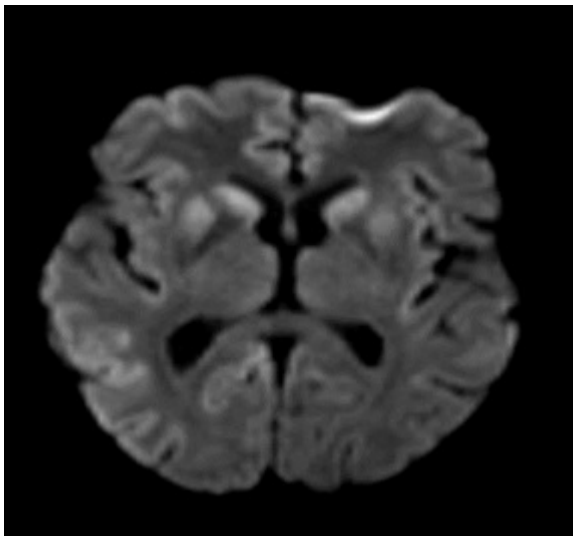

Figure 1: diffusion restriction of bilateral striatum and scattered area at right tempo occipital region and bilateral cingulate cortices resembling ribbon pattern.

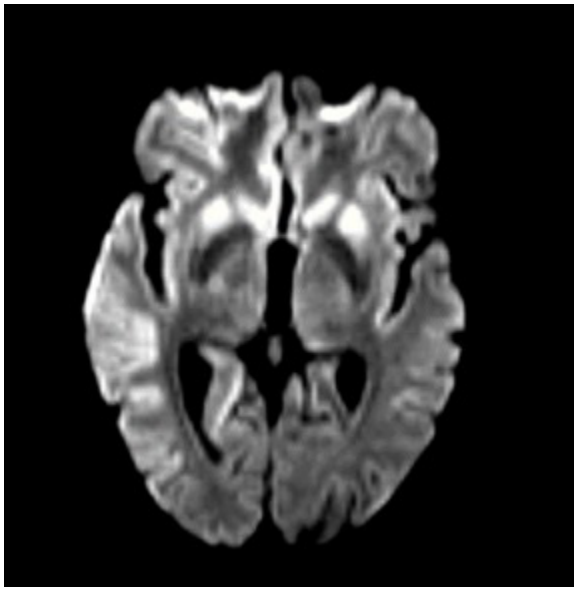

Figure 2: Diffusion restriction of bilateral striatum and scattered area at right tempo occipital region and bilateral cingulate cortices resembling ribbon pattern.

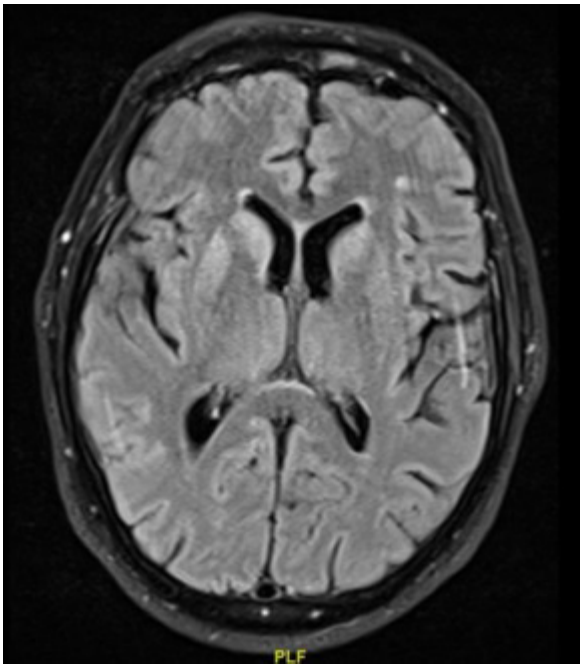

Figure 3: MRI brain showed fluid attenuated inverted recovery (FLAIR) signal abnormality of bilateral striatum and scattered area at right tempo occipital region and bilateral cingulate cortices resembling ribbon pattern.

3. Conclusion

CJD can have atypical clinical and radiological presentation. Diffuse epileptic form discharge (NCSE) on EEG in a patient with unexplained rapid cognitive decline and confusion might be a presentation of sCJD. Reversible causes of rapidly progressive dementia such as autoimmune, infectious, and toxic- metabolic etiologies must be excluded before making the final diagnosis of prion disease. Continuous video EEG monitoring is important, especially non convulsive status epilepticus is suspected [7]. Due to the fate of sCJD patients early suspicion need early detailed work up and complete expanded investigation so we can detect it early and avoid misdiagnosing such case.

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