We describe a rare case of a previously fit patient who presented with a rapidly progressive dementia and gait ataxia. EEG showed features of non-convulsive status epilepticus that was not responsive to anti-epileptic drugs. Post mortem brain biopsy confirmed sporadic Creutzfeldt-Jakob disease (sCJD).

A diagnosis of sCJD should be considered in patients with rapidly progressive dementia with non-convulsive status epilepticus especially with positive CSF protein 14-3-3 markers.

**Introduction**

Sporadic Creutzfeldt-Jakob disease (sCJD) is an insidious prion disease commonly presenting with rapidly progressive dementia, motor dysfunction, myoclonus, and characteristic periodic complexes on electroencephalogram (EEG). Partial, complex partial and secondary generalised seizures may occur in sCJD but are relatively uncommon, and status epilepticus is a rarity and may cause diagnostic confusion. Non convulsive status epilepticus (NCSE) is characterised by continuous epileptiform patterns on EEG, alteration of consciousness including coma and the absence of convulsive motor activity. We present an unusual patient who was admitted with a rapidly progressive global cognitive impairment, fluctuating level of consciousness and EEG features of NCSE. Post mortem examination of the brain confirmed sCJD.

**Case report**

A 63-year-old previously well right-handed housewife was admitted with a 1-month history of progressive unsteadiness of gait and changes in personality and behaviour. She had become apathetic, withdrawn, disoriented in time and unconcerned about her appearance. She had been unable to cope with her housework and increasingly dependent on her husband. Her verbal output had been reduced with inappropriate answers and she had been unable to follow conversations. She had a past history of hypothyroidism and took regular thyroxine replacement.

Initial neurological examination revealed receptive dysphasia. She was disoriented in time and place. Cranial nerves were intact. There was a bilateral Gegenhalten increase in muscle tone and motor perseverations but no grasp reflex. All reflexes were moderately brisk with flexor plantar responses. There was no myoclonus. Her gait was ataxic but a detailed assessment of limb coordination was not possible.

Routine blood and urine investigations, thyroid function and anti-thyroid antibodies, extensive autoimmune profile, anti-neuronal antibodies, serum electrophoresis, Vitamin B12 and folate levels and chest X-ray were normal. CT scan of the brain and the CSF examination were normal. However, the EEG revealed widespread, almost continuous repetitive 1-1.5Hz triphasic sharp and slow waves at 100-150uV, with an anterior emphasis, intermixed with rhythmic 5-7Hz theta waves in the posterior and central regions (20-30uV) (Figure 1) which did not alter during periods of agitation or eye opening. There was a brief response to intra-venous lorazepam. The EEG changes were suggestive of non-convulsive status epilepticus (NCSE). MRI scan of the brain revealed areas of high signal in the caudate and lentiform nuclei with normal signal in the thalamus, putamen, and corona radiata.

Figure 1: Almost continuous repetitive 1-1.5 Hz triphasic sharp, slow waves with an amplitude of 100-150uV that was widespread with an anterior emphasis intermixed with rhythmic 5-7 Hz theta waves in the posterior and central regions (20-30uV).
thalami. In view of these findings CSF was re-
examined for CJD markers was and was found to be positive for protein 14-3-3. 
The patient’s level of consciousness fluctuated widely with Glasgow Coma Score (GCS) of 5-
13. Repeated EEG’s showed her to be in persist-
tent NCSE that was resistant to treatment with 
phenytoin, sodium valproate, lorazepam and 
phenobarbitaline. 
Within days of admission the patient became 
bed-bound with increasing rigidity of all limbs, 
hyperorreflexia, incontinence and monosyllabic 
speech gradually progressing to akinetic 
musitis. Two weeks after admission, she deteri-
orated dramatically with a drop in GCS to 5 
accompanied by decorticate posturing. Repeat 
EEG confirmed her to be in NCSE with less than 
5 sec bursts of normal activity in spite of thera-
peutic serum phenytoin levels. She was aggres-
sively managed in the ITU with anaesthetic 
doses of propofol (achieving burst suppression), 
but remained in NCSE. She developed a chest 
infection and septicaemia and died a month after 
admission.
A post mortem brain examination revealed 
spongiiform change in the brain with neuronal 
loss in the cortex, deep grey nuclei and cerebel-
 lul with synaptic pattern of PrP immunopositi-
vity in the cortex, grey nuclei and cerebellum 
confirming sCJD.

Discussion
In 1920-21 Hans Jakob1 and Alfons Creutzfeldt2 first reported cases of a progressive dementing 
neurological disease which bears their name. This 
uncommon disease presents as a rapidly 
progressive dementia associated with a range of 
neurological signs, most commonly myoclonus of 
the limbs, cerebellar ataxia, and rigidity3 with 
a reported incidence of 1 per million-popula-
tion year.4 In the absence of neuropathology, the 
patients are classified as probable if they present 
with progressive dementia, typical EEG changes 
and at least two of the following: myoclonus, 
visual or cerebellar signs, pyramidal or extra-
pyramidal signs, akinetic mutism (Masters clas-
sification).5

Usual EEG changes in sporadic CJD include 
slowing down of background rhythms with 
periodic sharp wave complexes that may be lat-
eralized or diffuse6 and may or may not be syn-
chronous with myoclonus.7 Generalized or focal 
seizures have been known to occur in 10-15% 
patients with CJD in the course of the disease 
and are often resistant to anti epileptic drugs.8 
NCSE is characterised by continuous epilepti-
form patterns on EEG in a particular pattern 
(focal, general, or bi-hemispheric pattern), 
alteration of consciousness including coma and 
the absence of convulsive motor activity.9 
The MRI findings in our patient were typical 
of patients with sCJD as reported by 
Finkenstaedt et al. in 199610 with increased sig-
nal in bilateral caudate and putamen regions. 
The ‘pulvinar sign’ characterised by signal inten-
sity in the posterior thalamus commonly noted 
in variant CJD (vCJD) was not noted in our 
patient.11 Although the presence of protein 14-3-3 in 
the CSF indicates rapid brain destruction, its 
presence and persistence is more indicative of 
CJD as compared to other dementias or inflam-
matory processes in the brain.12 Furthermore, 
the presence of the protein is more indicative of 
sCJD13 although it may be raised in 50% of the 
patients with vCJD.14

Our patient was unusual in that, in addition to 
a rapidly progressive dementia and motor 
disturbances, she presented with features of 
NCSE, in the absence of a previous history of 
epilepsy. Indeed only a few cases have been 
reported with CJD presenting as NCSE.1415 
Schwinn et al.16 presented 4 patients diagnosed 
retrospectively with a mean age of 64 years pre-
senting with acute and sub acute changes in 
their mental status and NCSE. All were aggres-
sively treated with anti-convulsants and none 
showed clinical improvement (despite improve-
ment in their EEGs) with death ensuing within 
3 months.

Shapiro et al17 has recently reported a 70-year-
old patient with sCJD presenting with a one 
month history of deteriorating mental status, 
EEG revealed NCSE, which did not respond to 
standard anti-epileptic medication, and thus the 
patient was managed with a midazolam infu-
sion in the ICU in a bid to control the NCSE. 
The clinical description is very similar to our 
patient.

Conclusion
This case demonstrates the diagnostic dilemma 
when presented with a patient with progressive 
alteration in mental status and altered level of 
consciousness with no convulsive motor move-
ments. When the aetiology of NCSE cannot be 
established the diagnosis of CJD should be kept 
in mind. Serial EEGs, MRI scans and CSF eval-
uation for protein 14-3-3 can aid the clinical 
diagnosis and prognosis in these patients. 
Diagnosing CJD has significant consequences 
for the patient and potential risks for iatrogenic 
transmission need to be considered.

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