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CASE REPORT

Status Epilepticus Revealing Creutzfeldt-Jakob Disease: A Case Report

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ABSTRACT:
Creutzfeldt-Jakob disease (CJD) is a rare neurodegenerative disease of undetermined etiology, In Creutzfeldt-Jakob syndrome the symptomatology is quite variable and it consist of predominantly progressive dementia with a rapid onset, myoclonus, and also cerebellar, pyramidal, extrapyramidal and visual signs, the evolution of this disease is uniformly fatal, most patients die within 12 months, we present a case of a 72 year old women, who was admitted for a status epilepticus, She was later diagnosed with sporadic CJD.

Keywords: Creutzfeldt-Jakob, status epilepticus, case report

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Introduction:

Creutzfeldt-Jakob disease (CJD) is a rare neurodegenerative disease of undetermined etiology, responsible of a rapidly progressive dementia syndrome, usually the initial diagnosis of CJD can be obscured by its variable presentation. CJD poses a potential risk of iatrogenic transmission as it can incubate silently in humans for decades before becoming clinically apparent. There are three major groups of human prion disease: Sporadic, Genetic and Acquired. We present here a case of a 72-year-old woman, in whom the diagnosis of Creutzfeldt-Jakob disease was retained, on the basis of her clinical features, cerebrospinal fluid study, electroencephalogram and magnetic resonance imaging (MRI) sequences.

Case Presentation:

It is about Mrs B.F aged 72 years, referred to our medical intensive care unit of the university hospital for a state of epileptic seizure, to note that the patient has no particular medical or surgical history, in particular no psychiatric history nor the notion of dementia or other neurological disorders in the family. The history of the disease goes back to 03 months before her admission by the emergence of a progressive cognitive decline, change of behavior and personality, she was first noted to be aggressive and stopped taking care of herself. The evolution was marked by the appearance of generalized tonico-clonic seizures with no recovery of consciousness, which motivated the family to consult urgently, and then refered to us for specialized care.

The clinical examination on admission to the intensive care unit found an unconscious patient, Glasgow score of 11/15, with symmetrical and reactive pupils, without sensory-motor deficit, in addition the patient was hemodynamically and respiratory stable, blood pressure at 130/70mmH, heart rate at 92 beats per minute, recoloration time inferior to 03 seconds, diuresis preserved, SpO2 at 98% in the open air, respiratory rate at 19 cycles per minutes, temperature at 37. 5°C and capillary blood glucose was 1.5 g/l.

Due to the onset of convulsions with post-critical coma, the patient required orotracheal intubation and subsequent mechanical ventilation with an objective of RASS-5 sedation scale using midazolam 05mg/hour, after having received a loading dose of phenobarbital 10mg/kg.

Biological examinations requested on admission of the patient showed a natremia at 138 mmol/L, magnesemia at 1mmol/L, a calcemia at 2.41 mmol/L, urea at 5. 8 mmol/L, serum creatinine was 80 µmol/L, ASAT 35Ui/L, ALAT 28 Ul/L, hepatitis, retroviral and syphilitic serologies were negative, vitamin B9 and B12 dosage were normal, as well as the autoimmune disease workup in addition to TSH was unremarkable.

A chest X-ray and an abdominal-pelvic CT scan were requested without positive results.

An EEG showed periodic complexes and a generalized slowing down of the wave typical of Creudzfelt Jakob disease (figure 1).

Brain MRI showed symmetrical and bilateral signal abnormalities of the caudate and lenticular nuclei with abnormalities of the frontotemporal cortex (figure 2).

The research of protein 14-3-3 in the cerebral spinal fluid came back positive at 04 ng/mL, to note that the biochemical and cytological study of the CSF were normal with a proteinorachy at 0.4 g/l, a glycorachy at 0.5 and a ratio at 0.9.

The patient was treated with a triple therapy of phenobarbital, sodium valproate and levetiracetam. Despite this therapy, the patient convulsed as soon as the sedation was stopped.
The patient died with cerebral oedema complicated by supra-refractory convulsions.

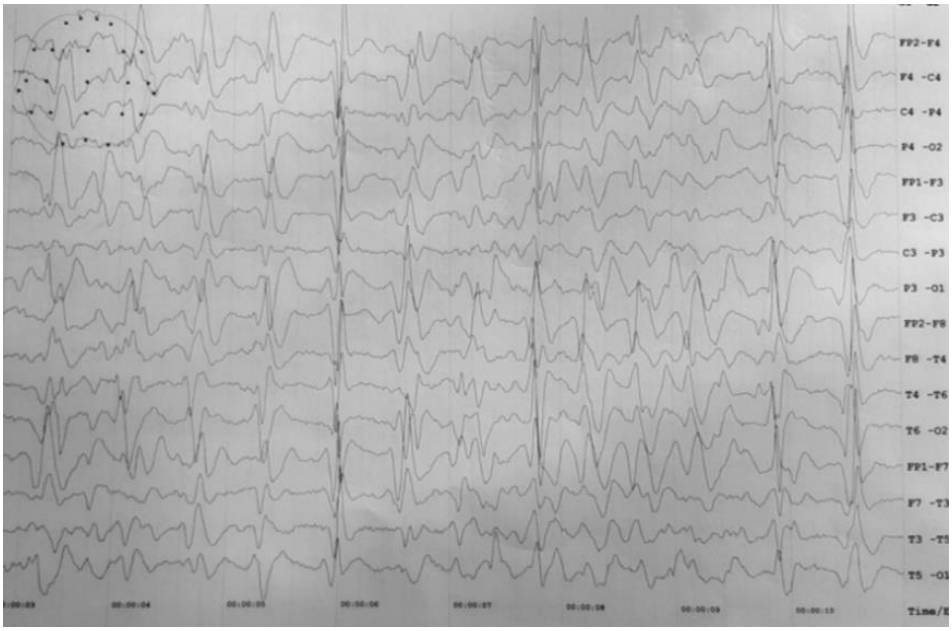

Figure 1: Electroencephalography characteristic of continuous periodic complexes and generalized slow waves

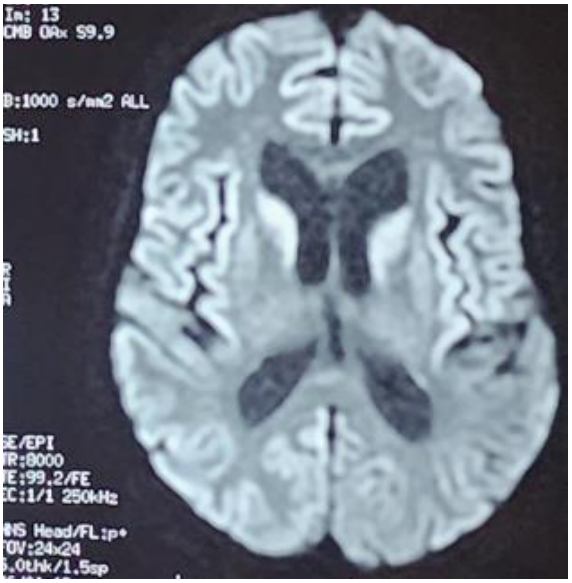

Figure 2: Brain MRI showing a symmetrical and bilateral hypersignal of the caudate and lenticular nuclei with anomaly of the frontotemporal cortex signal

Discussion:

Creutzfeldt Jakob syndrome is a spongiform neurodegenerative disorder, it is responsible for a subacute spongiform encephalopathy that can affect the central nervous system of humans, not to mention cases described in animals. The causative agent is not identified with certainty but, in all cases, it is observed the accumulation of the scrapie prion protein (PrPsc), abnormal form of the cellular prion protein (PrPc) [1].

It is a rare disease that affects 1 to 2 cases in 1 million, with an average age of 60.7 [2].

There are several etiologies of Creutzfeldt Jakob syndrome, the sporadic form represents the most common, followed by genetic forms, the acquired forms remain quite rare [3], in our case we suspect the sporadic origin of CZJ disease, based on the diagnostic criteria established by the CJD surveillance committee in 2017.Figure 1 [4].

Diagnostic criteria for surveillance of sporadic Creutzfeldt-Jakob disease from 1 January 2017.

1.1	DEFINITE:	
	Progressive neurological syndrome AND	
	Neuropathologically or immunohistochemically or biochemically confirmed	
1.2	PROBABLE:	
	1.2.1	I + two of II and typical electroencephalogram [a]
OR	1.2.2	I + two of II and typical magnetic resonance imaging brain scan [b]
OR	1.2.3	I + two of II and positive cerebrospinal fluid (CSF) 14-3-3
OR	1.2.4	Progressive neurological syndrome and positive real-time quaking-induced conversion in CSF or other tissues
1.3	POSSIBLE:	
	I + two of II + duration <2 years	
I	Rapidly progressive cognitive impairment	
II	A	Myoclonus
	B	Visual or cerebellar problems
	C	Pyramidal or extrapyramidal features
	D	Akinetic mutism

Figure 3: Diagnostic criteria for sporadic CJD

The latency period may last several years; however, death usually occurs within the first year after the onset of symptoms.

In Creutzfeldt-Jakob syndrome the symptomatology is quite variable, there are no specific symptoms, but in the majority of cases, the symptomatology starts with psychiatric-like disorders, such as cognitive decline rapidly progressing to dementia[5], after the psychiatric-like symptoms, Neurological signs are found that translate the suffering of the central nervous system, such as myoclonus, cerebellar ataxia and status epilepticus[6], the most prominent of which are changes in behaviour, emotional response and intellectual function, together with abnormalities of cerebellar function[7] in our case, the patient presented a dementia syndrome neglected by the family and which evolved rapidly to a status epilepticus.

As far as the paraclinical examinations are concerned, the electroencephalogram can help to suspect the diagnosis, initially, the tracing is often slowed down, and then we note the presence of polymorphic and periodic slow wave discharges with paroxysmal bi- or triphasic peak-waves [8], The typical duration of these waves is 100–600 ms, recurring every 0.5–2 s[9].

The analysis of the radiological semiology of encephalic magnetic resonance imaging (MRI) can bring elements that can be in favor of CJD, such as hypersignals localized in the caudate nucleus, putamen or on the cerebral and cerebellar cortex, to note that the FLAIR sequence allows better visualization of these lesions[10] , which is consistent with our case.

The biochemical and bacteriological examination of the CSF is often normal, but the presence of Tau or 14-3-3 proteins in the CSF has an interesting diagnostic value[11].

Diagnostic certainty is only obtained by histological study of the brain tissue, most often post mortem. A positive examination reveals the characteristic lesion association: spongiosa, neuronal rarefaction, astrocytic gliosis and sometimes amyloid plaques [12].

We can suspect CJD by a clinical diagnostic criteria use a combination of characteristic neuropsychiatric symptoms, CSF proteins 14-3-3, MRI, and EEG[13].

Conclusion:

Creutzfeldt Jakob Syndrome is a neurological syndrome characterized by its clinical polymorphism, its rarity as well as the complexity of its diagnosis. Nevertheless, and through our case, CJS must be suspected in front of a status epilepticus refractory to all therapeutic tools, and after having ruled out organic, metabolic, infectious or autoimmune etiologies.

Conflict of interest:

The authors report no conflict of interests

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