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Case Report

The First Case of Creutzfeldt - Jakob Disease in Mongolia

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Abstract

Creutzfeldt-Jakob disease (CJD) is a spongiform encephalopathy that results in a rapidly progressive dementia. This disease is uniformly fatal, with the majority of patients succumbing within a year or less from the onset. Although the vast majority of cases are sporadic, occasional instances of familial and acquired forms are also encountered. Clinical presentations involve rapidly progressive cognitive impairment, myoclonus, visual disturbances, and a range of non-specific signs including cerebellar, pyramidal, and extrapyramidal symptoms. The challenge in diagnosing CJD stems from the non-specific nature of these findings and the difficulty associated with awareness and clinical suspicion, making the diagnostic process often intricate and demanding. We present a 65-year-old man with a 5-month history of rapidly progressive dementia. After a series of diagnostic examinations and continuous follow-up, he was diagnosed with probable sporadic Creutzfeldt-Jakob disease based on Centers for Disease Control and Prevention (CDC) in 2018 criteria for the Diagnosis of Sporadic CJD with key findings of rapidly progressive dementia, extrapyramidal signs, and abnormal hyperintensity signals on diffusion-weighted MRI, and typical periodic sharp wave complexes on EEG. His symptoms progressively worsened and was developed a state of akinetic mutism 3 months after the onset. A literature review was performed on differential diagnoses that present with rapidly progressive dementia and thereby mimic sporadic Creutzfeldt-Jakob disease.

Keywords: Sporadic Creutzfeldt - Jakob Disease; Prion Disease; Rapidly Progressing Dementia

Introduction

Creutzfeldt-Jakob Disease (CJD) or subacute spongiform encephalopathy is a rapidly progressive human prion neurodegenerative disorder believed to be caused by an abnormal isoform of a cellular glycoprotein known as the misfolded prion protein [1]. There are currently four major clinical forms of prion disease in humans and animals (Creutzfeldt-Jakob disease, kuru, Gerstmann-Straussler-Scheinker syndrome, and fatal insomnia) [1,2]. Prion protein differs from viruses in that it lacks nucleic acid, does not have a specific structure, and is usually found on the surface of brain cells (encoded by a gene on chromosome 20) in a healthy human body [2]. Misfolded prion proteins enter the brain cells through inheritance or external infection which ultimately results in damage to brain tissue and replaced by glial growth or amyloid-containing plaques to the development of characteristic microscopic holes, giving the brain a spongy appearance primarily observable in regions such as the cerebral cortex, thalamus, striatum and cerebellum [3].

CJD occurs worldwide and the estimated annual incidence in many countries, has been reported to be about 1 case per million population [4]. CJD was first described by the German scientist Hans Gerhard Creutzfeldt in 1920, and soon after Alfons Maria Jakob confirmed it in another study in 1921, it was named Creutzfeldt-Jakob disease [1,2].

There are four different forms of CJD that have been identified: familial, iatrogenic, variant, and sporadic. The incubation period after infection with CJD lasts 2-20 years and the sporadic form accounts for 85%-90%, while the remaining 10-15% are familial, iatrogenic forms, and variants [5]. The sporadic case occurs at the age of 55-70, familial at the age of 40-55, the new variant occurs at the age of less than 50, and the average age of the disease is 65, but the disease is not registered at the age of 30 and over 80, and the gender ratio does not differ [6].

The familial form of CJD is triggered by a mutation in the gene encoding the prion protein (PRNP) at codons 178, 200, or 210, leading to the formation of abnormal PrP protein [2,3]. Conversely, the sporadic form results from the spontaneous transformation of PrPc protein into PrPSc protein or the emergence of mutations in the PrP gene and the amino acid sequence of the prion protein isolated from the patient remains identical to that of the normal PrPc protein isoform [2,3,6]. Molecular-genetic studies have shown that hypersensitivity to prion protein is associated with a 129-codon PrP protein gene polymorphism that encodes valine and methionine [6]. The iatrogenic variant arises from the homozygosity for valine at codon 129 of the PrP, typically associated with the use of growth hormone [3,4]. In contrast, the sporadic variant is linked to homozygosity for methionine at codon 129 in 78% of patients (compared to 48% in the general population) [3,4]. The occurrence of heterozygotes for methionine and valine is notably infrequent in comparison to the control group (12% and 42%) [3-5]. This observation suggests a potential genetic predisposition associated with the variant in question [3,4].

Clinical findings include rapidly progressive cognitive impairment, myoclonus, visual disturbances, cerebellar, pyramidal, and extrapyramidal signs [2,3,6]. At the final stages of disease, there's a progression towards akinetic mutism, culminating in eventual fatality. These signs are nonspecific and can be caused by a variety of diseases that manifest as progressive dementia [1-3,6]. This disease is uniformly fatal and most patients die within a year from onset [1,4,5]. Although the diagnosis of CJD is confirmed by the detection of abnormal prion proteins in the brain, Periodic Sharp-Wave Complexes (PSWC) in the Electroencephalogram (EEG), Diffusion Weighted Imaging (DWI), Fluid Attenuated Inversion Recovery (FLAIR) signal hyperintensities around the putamen, caudate and 14-3-3 protein in the Cerebrospinal Fluid (CSF) are quite possible to confirm the diagnosis [7]. In most cases, the patient dies within 6 months to 1-2 years and there is no effective treatment.

We provide a case report featuring the initial occurrence of Sporadic Creutzfeldt-Jakob Disease (sCJD) in Mongolia. Additionally, we highlight the intricacies associated with diagnosing this disease during its early stages in clinical courses within resource-limited settings.

Case Report

A 65-year-old male, employed as a water engineer and fruit farmer in a rural area of Mongolia, sought medical attention due to a 2-month history of behavioral and personality changes, coupled with a noticeable decline in cognitive function. As reported by the family, the individual started experiencing memory problems related to recent events and exhibited confusion and within 2 weeks, his cognitive functions were profoundly impaired. He consistently took antihypertensive medications to manage hypertension and occasionally consumed alcohol, as reported. There is no recorded history of psychiatric issues, prior surgeries, blood transfusions, or hormonal injections for him. He was treated in the neurology department and intensive care unit of the First, Third Central Hospitals, National Psychiatry Center and according to the medical documents, several diagnoses such as "Vascular Encephalopathy", "Vascular dementia", "Epilepsy and Status of Epilepticus" were made.

On initial neuroradiological findings, the patient showed signs of being inactive in their surroundings, responded to questions with inaccuracies in single-word answers, stated his age incorrectly, lacked awareness of the current date and time, failed to recognize their own children, struggled with the left-hand to right-ear test, had difficulty maintaining left-right orientation, was unable to perform numerical operations within a ten-unit range, and his name was misspelled. The patient presented with aphasia, which affects their ability to express and understand language (motor and sensor aphasia). There was also a lack of action, known as apraxia, which impairs their ability to carry out purposeful movements. Moreover, he had experienced a lack of recognition, referred to as agnosia, leading to difficulty in identifying familiar objects or people. Additionally, the individual showed an inability to meet the summary criteria for assessing Mini Mental State Examination (MMSE), possibly indicating cognitive impairment. There was a slight increase in muscle tone and tendon reflexes increased. Rossolimo and Hoffmann reflexes in the hands were present, while mouth-lip automatism remained evident. He was unable to perform toe-nose and heel-knee-shin tests, suggesting coordination issues. Positive Romberg test and Tandem gait was mildly unstable. The ultrasound examination revealed no abnormalities or changes in the internal organs. Furthermore, the patient's blood and biochemical parameters were within the normal range based on the assessment. The EEG showed a suppression of basal electrical activity in all deprivations, along with generalized epileptic paroxysms. In non-contrast MRI-DWI imaging there was increased signal intensity (hyperintensity) in the parietal, frontal, temporal lobe cortex,

basal ganglia, and left hemisphere of the cerebellum. The clinical features, progression of the disease course, EEG, and findings of brain imaging led us to the diagnosis of probable Creutzfeldt–Jakob disease (spongiform encephalopathy) based on Centers for Disease Control and Prevention (CDC) in 2018 criteria for the Diagnosis of Sporadic CJD with key findings of rapidly progressive dementia, extrapyramidal signs, and asymmetric hyperintensity signals in the bilateral parietal, frontal and temporal lobes, and basal ganglia on diffusion-weighted MRI. Our management protocol consisted of symptomatic treatment including seizure control (with a 3-week course of sodium valproate) and strictly adhering to an infection prevention regimen.

3 months after the onset of the disease, when the patient was being treated in the intensive care unit of the First Central Hospital, during the second examination by our consulting neurologist, as noted in the medical history, the patient came with unconsciousness and permanent myoclonus convulsions. During the neurological examination, since previous examination, he was very emaciated, unable to communicate verbally, responded to pain stimuli (pressing on the upper edge of the back, pinching the tips of the fingers), opening his eyes, grimacing, slightly moving his arms and legs, muscle tone was increased, tendon reflexes were weak, Hoffmann and Rossolimo reflexes in the fingers, pronounced mouth-lip automatism, Babinski sign were present on the left side. There were no changes in cardiovascular, respiratory, and vital functions; blood (CBC), biochemistry, cerebrospinal fluid (cell count, protein, glucose) and syphilis and HIV tests were unremarkable. In the EEG Bilateral Periodic Epileptiform Discharges (BiPEDs) were noted (Figure 1). In the MRI-DWI axial imaging of the brain, focal changes (more on the left) with increased signal intensity (more on the left) were detected in parietal, frontal, temporal cortex, cingulate girus, caudate and putamen (Figure 2). These changes significantly increased from the changes in previous MRI and patient under the ccontinuous use of anticonvulsant drugs (Valproate 1200 mg per day). “Probable Sporadic Creutzfeldt-Jakob disease, spongiform encephalopathy” was diagnosed, based on CDC’s Diagnostic Criteria for Sporadic Creutzfeldt-Jakob Disease (sCJD), 2018 with key findings of rapidly progressive dementia, extrapyramidal signs, and asymmetric hyperintensity signals in the bilateral parietal, frontal and temporal lobes, and basal ganglia on diffusion-weighted MRI, and typical periodic sharp wave complexes on EEG. The patient was advised to continue symptomatic treatment under medical supervision and strictly adhere to the disinfection regimen. The patient’s general condition had rapidly deteriorated and sent to the rural hospital where he was kept comfortable and passed away while at home after a couple of months.

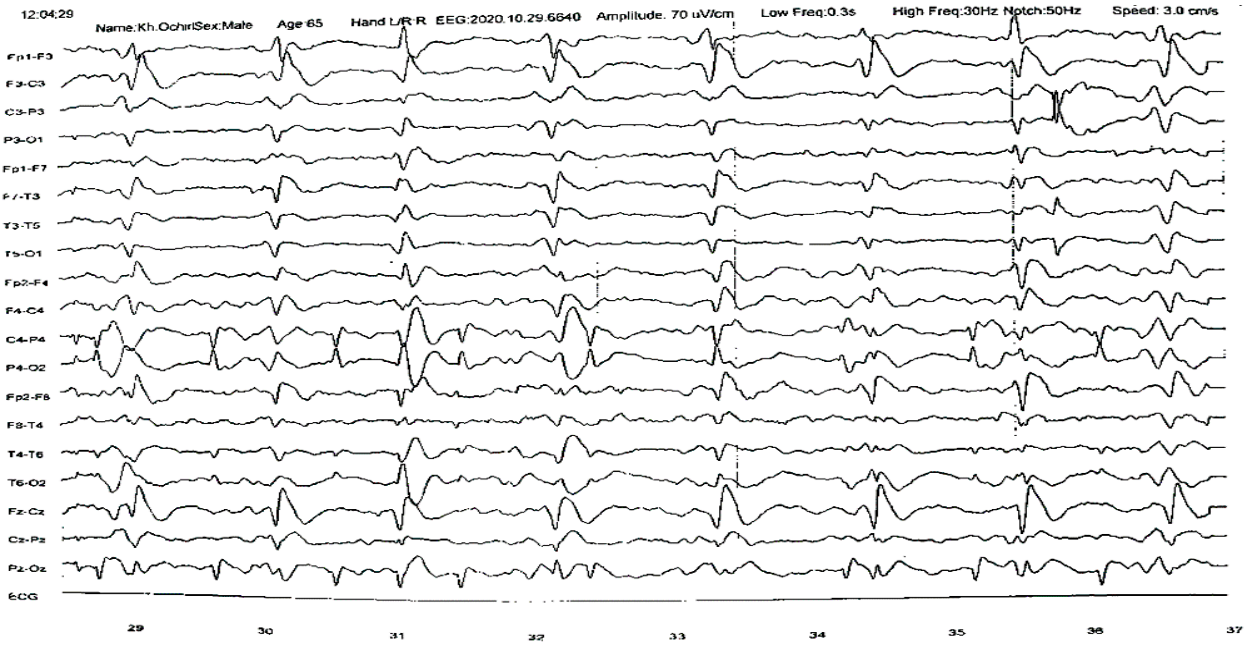

Figure 1: Patient H.O. 65 years old, male, diagnosis: sCJD. EEG: bilateral periodic epileptiform discharges (BiPEDs).

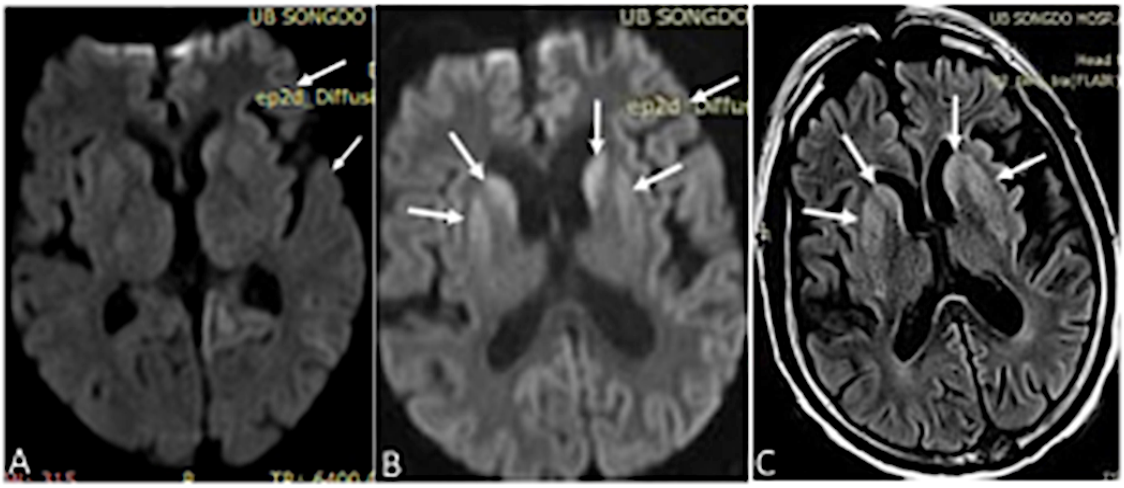

Figure 2: Patient H.O. 65 years old, male, diagnosis: sCJD; In the MRI-DWI axial imaging of the brain, focal changes (more on the left) with increased signal intensity (more on the left) were detected in (A) parietal, frontal, temporal cortex, cingulate girus, (B, C) caudate and putamen.

MRI images of this patient's brain were obtained from The National Prion Disease Pathology Surveillance Center, Department of Neuroradiology, and submitted an opinion confirming our diagnosis. Based on preliminary results of an ongoing study with MRI of 770 autopsy confirmed patients referred to the NPDPSC, the sensitivity, specificity and diagnostic accuracy of MRI (with DWI and/or FLAIR) in the diagnosis of sCJD is 93,05%, 96,69% and 93,77%, respectively [8].

Discussion

Although the diagnosis of CJD is confirmed by the detection of abnormal prion proteins in the brain, Periodic Sharp-Wave Complexes (PSWC) in the EEG, DWI, FLAIR signal hyperintensities around the putamen, caudate and 14-3-3 protein in the cerebrospinal fluid are quite possible to confirm the diagnosis [1,4,5]. Dementia, myoclonus, visual disturbances, ataxia, pyramidal and extrapyramidal signs on neurological examination do not occur selectively in every clinical scenario of CJD, and these non-specific symptoms are observed in a number of neurodegenerative diseases, therefore are important in differentiating the diagnosis from other disorders.

According to the latest diagnostic criteria released by the Centers for Disease Control and Prevention (CDC) in 2018, our case qualifies as a presentation of sCJD [7]. Furthermore, within a brief span of 2-3 weeks, the individual exhibited rapid onset of dementia and initially indications of cerebellar ataxia, followed by the emergence of pyramidal and extrapyramidal alterations, culminating in manifestations of akinetic mutism. The condition has advanced over time which eventually leads to myoclonus, EEG recordings displayed periodic bilateral periodic epileptiform discharges (BiPEDs) (Figure. 1) were selectively evident. Additionally, DWI illustrated focal changes in depth within the bilateral fronto-occipital region, caudate, and basal ganglia (Figure 2). These findings collectively affirm the classification of this case as a form of CJD.

CJD is very rare, however mimics are common and there is a necessity to distinguish among diseases that manifest with rapidly progressive dementia including Alzheimer's disease. Levy body dementia, Frontotemporal Dementia (FTD) and corticobasal degeneration. Alzheimer's disease can be confused with CJD in cases of rapid progression, myoclonus, periodic sharp waves in EEG, and positive 14-3-3 protein in CSF [9,10]. However, cognitive decline can progress over several years and last for 10 years or more [10]. During the course of the disease, a person's behavior changes, language, work behavior, and experience are lost, which will slowly lead to functional disability. However, in the case of Alzheimer's disease, head CT and MRI scans show signs of cerebral atrophy, especially around the hippocampus and cortex, while in the case of CJD, the atrophy is small or not at all [9,10]. On the other hand, in the case of CJD, MRI analysis reveals a characteristic increase in signal intensity in the bilateral caudate nucleus and corpus callosum [9]. Levy body dementia is also characterized by progressive dementia and cognitive changes, accompanied by visual hallucinations, Parkinsonism, and myoclonus [11]. However, when considering CJD, the symptoms of dementia gradually worsen, visual hallucinations are observed early in the course of the disease, and periodic sharp waves are sometimes seen on the EEG [11]. Although extrapyramidal

symptoms such as rigidity, tremor, and akinesia in FTD can mimic CJD, in most cases, frontal and/or temporal lobe atrophy is seen on MRI [12]. Corticobasal degeneration is a neurodegenerative disorder of uncertain origin that impacts the basal ganglia and cerebral cortex, particularly the frontal and parietal lobes [13]. This advancing condition typically gives rise to progressive dementia, myoclonus, hallucinations, the "alien hand" phenomenon, and Parkinsonism symptoms like bradykinesia, rigidity, or gait disturbances—all of which may also manifest in individuals with sCJD [13]. Although the age of onset for corticobasal degeneration aligns with that of sCJD at 60-80 years old, it's crucial to note that the progression of corticobasal degeneration is characterized by a less rapid and more protracted course and this condition typically exhibits a much longer duration, spanning 8-10 years [13].

Other differential diagnoses include meningoencephalitis, Paraneoplastic Encephalomyelitis (PNE), Progressive Supranuclear Palsy (PSP) and Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL). In clinical practice, the manifestation of conditioned meningoencephalitis can be attributed to a diverse array of causes [14]. These may include viral factors such as herpes virus, HIV, and the Japanese encephalitis virus, bacterial contributors like streptococcus and Lyme disease, fungal infections such as cryptococcus, parasitic agents including toxoplasma and malaria, and autoimmune factors exemplified by Hashimoto's encephalitis and limbic encephalitis [14]. Typically, meningoencephalitis initiates abruptly, marked by symptoms such as fever, headache, confusion, and epileptic seizures. Memory alterations exhibit a relatively swift pace of change when compared to conditions like CJD and other forms of dementia [14]. An essential characteristic of meningoencephalitis is the elevation in the number of cells and protein levels in the CSF, a feature absent in CJD [14]. Nevertheless, it's noteworthy that in instances of herpes virus-induced limbic encephalitis, similarities in memory changes and myoclonus seizures to those observed in CJD may be apparent [15]. Progressive Supranuclear Palsy (PSP) represents another neurodegenerative disorder marked by cognitive decline, vertical supranuclear ophthalmoplegia, visual disturbances, extrapyramidal signs, and gait disturbances leading to exclude from CJD [16]. Early manifestations include postural instability and limitations in vertical gaze [16]. Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is identified by progressive dementia, mood disorders, migraine headaches, and Transient Ischemic Attacks (TIAs) or strokes [17]. The predominant presentation often involves recurrent TIAs or strokes occurring around the age of 40-50, which is earlier than the onset of sCJD [17]. MRI scans reveal distinct multiple hyperintensities in the periventricular white matter of the frontal lobes [17]. The significance of family history is crucial, as CADASIL is an inherited disorder resulting from a mutation in the Notch 3 gene, and a positive family history is common among CADASIL patients [17]. Paraneoplastic Encephalomyelitis (PEM) is frequently linked to malignancies, with lung cancer being the most prevalent (80%) [18]. This condition can affect multiple regions of the nervous system and common clinical manifestations encompass subacute cognitive decline, alongside a range of neurological symptoms such as personality changes, depression, anxiety, hallucinations, agitation, and seizures [18]. The specific symptoms vary based on the affected areas of the brain. Cerebrospinal Fluid (CSF) analysis serves as a valuable tool in distinguishing between sCJD and PEM [18]. While the protein 14-3-3 can be identified in the CSF of both PEM and sCJD cases, inflammatory markers in the CSF are typically observed solely in PEM and not in sCJD and additionally, anti-Hu antibodies are often detected in cases of PEM [18].

Marchiafava-Bignami Disease (MBD) and Unverricht-Lundborg Disease (ULD) have features that distinguish them from CJD, but it's also important to note that some symptoms may overlap, leading to diagnostic challenges. MBD is a rare neurological disorder characterized by demyelination and necrosis of the corpus callosum, is often associated with chronic alcoholism, although non-alcoholic cases have been reported [19]. MBD typically presents with symptoms such as confusion, seizures, behavioral changes, and impaired movements [18]. The exact cause of MBD is not fully understood, but chronic alcohol abuse is considered a significant risk factor and malnutrition, vitamin B deficiency, and oxidative stress are also believed to contribute to the development of MBD [19]. Diagnosis of MBD involves brain imaging studies, such as Magnetic Resonance Imaging (MRI), which can reveal characteristic changes in the corpus callosum which is characteristic feature helps differentiate MBD from other neurological disorders, including CJD [19]. Unverricht-Lundborg Disease (ULD) typically manifests in the second decade of life, presenting with generalized forceful-tremor seizures and persistent myoclonic convulsions affecting both sides of the body [20]. Individuals with this condition may also experience muscle tone stiffness, progressive deepening of dementia symptoms, loss of balance, vertigo, and visual changes [20]. The disease follows an autosomal dominant inheritance pattern, meaning that a single copy of the mutated gene from either parent can lead to its expression [20]. The diagnosis of seizures in ULD often occurs at a relatively young age and is characterized by stable epileptic discharges on EEG [20]. This distinctive EEG pattern helps differentiate ULD from CJD, which may present with different EEG findings [20].

Conclusion

When diagnosing the first case of CJD in our country, we observed multifocal neurological symptoms, including the rapid progression of dementia, changes in balance, myoclonus, akinesia, and rigidity. Key diagnostic parameters included characteristic high-amplitude periodic sharp waves detected by EEG and MRI features. This comprehensive assessment, based on key neuroclinical, neurophysiological, and neuroradiological parameters, contributed to the accurate diagnosis of CJD. According to literature reports, EEG findings may not consistently exhibit a specific association of sharp waves in cases of new, growth hormone-dependent, medically induced forms of CJD. Therefore, in the last years, for the purpose of research and confirming the diagnosis of CJD, it is recommended to assess the presence of the 14-3-3 protein in the CSF. Following the diagnostic criteria outlined by the Centers for Disease Control and Prevention (CDC) in 2018 is also advisable. Additionally, the importance of employing supplementary analysis methods, including standard structural analysis of brain tissue, Misfolding Cyclic Amplification (PMCA) CSF analysis for total tau, as well as real-time quaking-induced conversion (RT-QuIC), has been underscored in the diagnostic process.

Due to the presence of abnormal prion proteins, cattle, sheep, deer, and antelope can be susceptible to prion diseases. Consequently, the consumption of under processed animal products, particularly brains and spinal cords, is prohibited to mitigate the risk of transmission. As our country relies significantly on livestock, there is an ongoing concern about the potential occurrence of prion diseases in the future. In the event of a prion disease, it becomes crucial to differentiate it from other treatable cognitive disorders at an early stage. This early distinction allows for prompt intervention and management. Equally important is the need to educate the affected individual's family about the nature of the disease and to strictly adhere to infection prevention procedures during caregiving. These precautions are essential due to the unique characteristics of prion diseases, their potential for transmission, and the lack of effective treatments. By fostering awareness, early diagnosis, and implementing strict preventive measures, it is possible to enhance the overall management and understanding of prion diseases in a livestock-oriented country.

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Conflict of Interest Statement: The authors declare that no conflicts of interest exist.

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