International Medical Case Reports Journal

Dovepress
Taylor & Francis Group

Open Access Full Text Article

CASE REPORT

First Suspected Case of Sporadic Creutzfeldt-Jakob Disease in Syria

Hamed Tarboosh[1], Raghed Mansour[1], Noor Haidar[2], Pierre Ghulam[1,3], Reem Gergy[1,3], Faten Nafla[3]

[1]Faculty of Medicine, Latakia University, Latakia, Latakia Governorate, Syrian Arab Republic; [2]Faculty of Pharmacy, Latakia University, Latakia, Latakia Governorate, Syrian Arab Republic; [3]Department of Neurology, Latakia University Hospital, Latakia, Latakia Governorate, Syrian Arab Republic

Correspondence: Hamed Tarboosh, Faculty of Medicine, Latakia University, PO Box 2230, Latakia, Latakia Governorate, Syrian Arab Republic, Email hamedtarboosh5@gmail.com

Video abstract

Point your SmartPhone at the code above. If you have a QR code reader the video abstract will appear. Or use: https://youtu.be/jDtv1LSw2r0

Abstract: Prion diseases are rare, fatal neurodegenerative disorders caused by misfolded proteins. Sporadic Creutzfeldt–Jakob Disease (sCJD) is the most common form and develops without any identifiable triggers. This case report documents the first probable diagnosis of sCJD in Syria, involving a 72-year-old woman who presented with depressive symptoms for several months before rapidly deteriorating, and eventually succumbing to complications of the disease. Clinical examination findings, exclusion of CJD-mimicking conditions, and specific MRI findings, supported the diagnosis in the absence of biochemical tests or postmortem confirmation. This report emphasizes the challenges of diagnosing prion diseases in resource-limited settings and calls for the development of national surveillance systems in low- and middle-income countries to aid early detection, monitor the global disease burden, and reduce the risk of outbreaks. Raising awareness of prion diseases among physicians in such contexts is crucial for improving disease recognition.

Plain Language Summary: This report describes the first likely case of a rare brain disease in Syria. The disease, called sporadic Creutzfeldt-Jakob Disease (sCJD), is caused by misfolded proteins and is always fatal. The patient was a 72-year-old woman. She first showed signs of depression for a few months. After that, her condition worsened very quickly, and she died from complications of the disease. Because specialized lab tests and an autopsy were not available, doctors diagnosed her based on her symptoms, by ruling out other diseases that look similar, and on specific brain scan (MRI) results. This case shows how hard it is to diagnose such diseases in places with limited medical resources. The report recommends that lower-income countries create national systems to track this disease. This would help find cases early, better understand how common it is worldwide, and prevent outbreaks. It is also crucial to train doctors in these areas to recognize the disease.

Keywords: epidemiology, neurology, prion diseases, rapidly progressive dementia

Introduction

Prion diseases are rare and fatal neurodegenerative diseases caused by the buildup of a misfolded prion protein, leading to neural apoptosis and spongiform degeneration of brain tissue. Sporadic Creutzfeldt–Jakob Disease (sCJD), which has an unknown cause, represents more than 85% of CJD cases, with an incidence of 1.5 to 2 per million worldwide annually.[1] However, a recent global epidemiology atlas indicates that formal surveillance data are only available from 34 countries worldwide Figure 1.[2] On the other hand, data from low- and middle-income countries (LMICs) primarily rely on case reports or case series.[3–9] This lack of data is particularly concerning for LMICs, where underreporting is common due to limited diagnostic capacity.[10] Less frequent forms include: genetic CJD (gCJD) due to inherited disease-causing mutations in the prion protein gene PRNP, iatrogenic CJD (iCJD) from medical procedures, and variant CJD (vCJD) from consuming contaminated beef.[1] CJD progresses rapidly, with nearly 90% of patients dying within a year of symptom onset. The clinical

Received: 26 October 2025
Accepted: 15 December 2025
Published: 22 December 2025

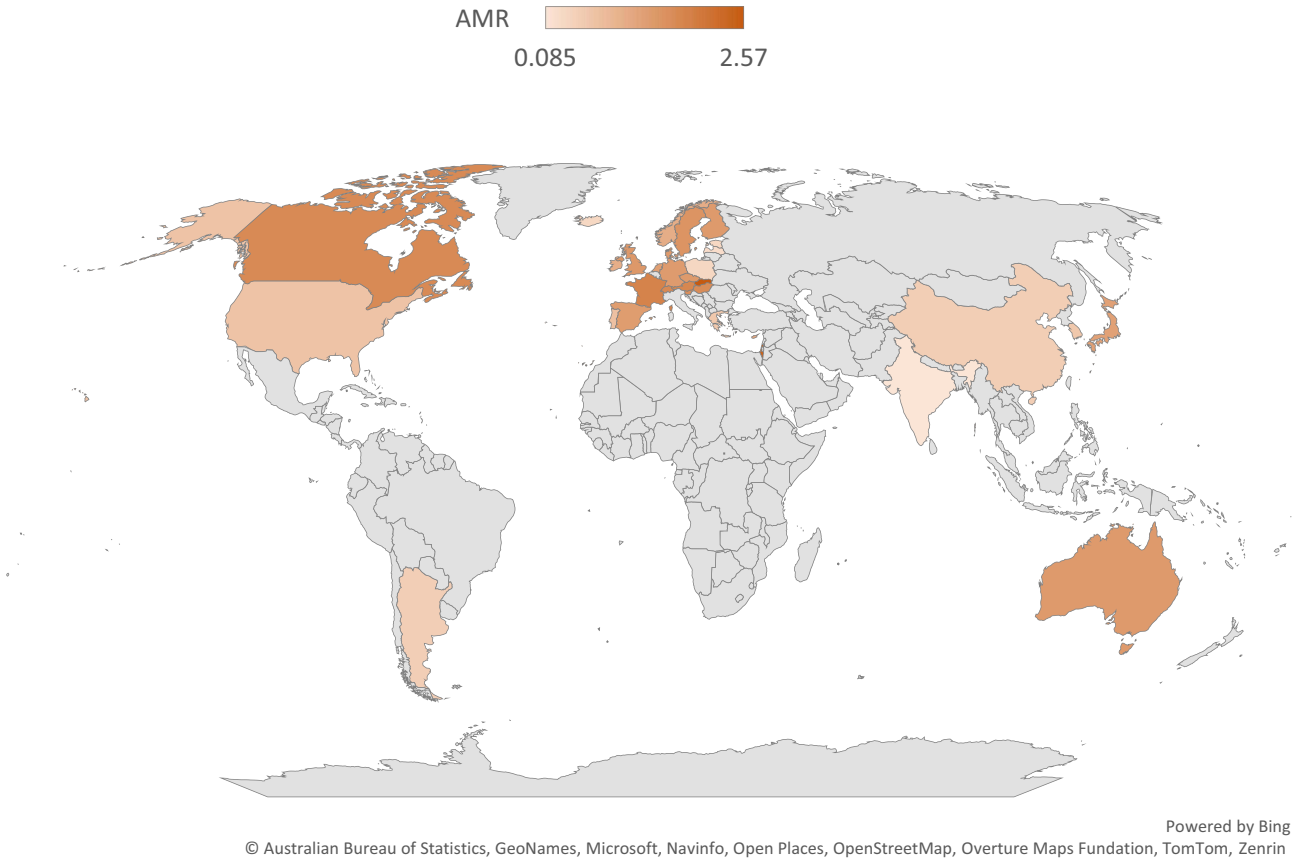

Figure 1 Heatmap showing the global annual mortality rate (AMR) of sporadic Creutzfeldt–Jakob Disease illustrated as color gradients (cases per million).

presentation is nonspecific and includes myoclonus, cognitive impairment, rapidly progressive dementia, hallucinations, and cerebellar signs.[11] While sCJD may not be initially suspected, it should be considered in cases of rapid cognitive decline. This paper discusses a case of probable sCJD, which is, to our knowledge, the first in Syria. We address diagnostic difficulties in low-income countries, where biochemical confirmation is costly, making clinical findings and neuroimaging crucial. We also highlight the need for surveillance protocols to document future cases and prevent outbreaks.

Case Report

History

A 72-year-old woman presented to our hospital after rapid deterioration of consciousness level and motor function. The patient was a life-long nonsmoker and abstained from alcohol. Medical history included hypertension. A detailed history was taken from family members and revealed inattention, failure to perform daily activities, apathy, disorientation to time and place, notable personality changes, and altered speech patterns starting two months before presentation Figure 2. The patient then developed visual hallucinations, behavioral abnormalities (placed her shoes in the refrigerator), and poorly defined involuntary movements that subsided during sleep. Notably, she exhibited depressive symptoms for several months preceding the neurocognitive decline. The patient was examined by several neurologists who excluded thyroid function abnormalities and vitamin B12 deficiency, however, the diagnosis remained inconclusive.

Admission

A local neurologist referred the patient to our hospital upon worsening consciousness level and motor function. Physical examination upon admission revealed positive Hoffman and Babinski signs bilaterally, and rigidity in all four limbs. Further testing revealed pyuria and blood granulocytosis. The patient was hospitalized for supportive care and case study,

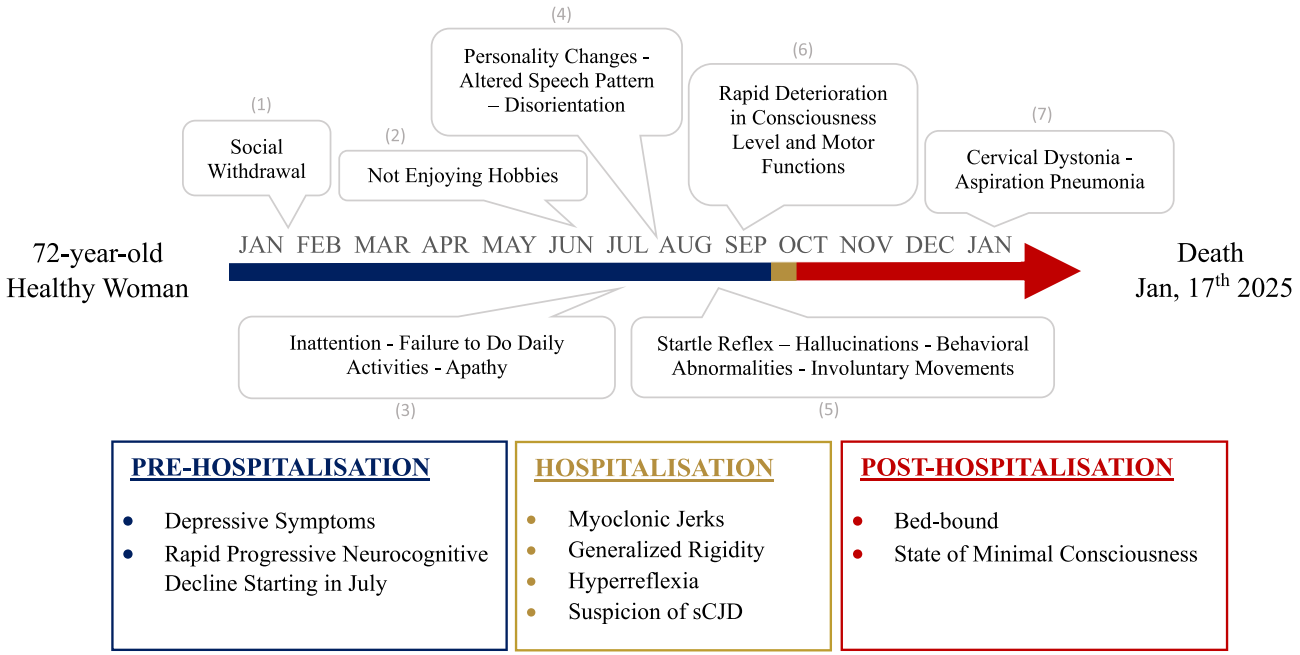

Figure 2 Timeline of the progression of the disease detailing major signs and symptoms divided into three periods: prehospitalisation, hospitalisation, and posthospitalisation.

during which she developed myoclonic jerks and hyperreflexia, and received a brain magnetic resonance imaging (MRI) scan from a nearby private facility showing multiple abnormalities. Table 1 and Figure 3 detail the patient's significant work-up results and key examination findings.

Diagnosis

Given the patient's history of rapidly progressive neuropsychiatric disorder, clinically evident myoclonus, and extrapyramidal and pyramidal signs, along with the characteristic brain MRI findings, a diagnosis of probable sCJD was established based on the CDC diagnostic criteria,[12] which require: (1) Progressive dementia; and (2) At least two of the following clinical features: myoclonus, visual or cerebellar signs, pyramidal/extrapyramidal signs, or akinetic mutism; along with (3) A typical MRI finding of high signal in the caudate nucleus and putamen or at least two cortical regions on Diffusion-Weighted Imaging (DWI) or Fluid-Attenuated Inversion Recovery (FLAIR). As noted in Figure 3, the DWI abnormalities are present in almost all cortical regions with thalamic and striatal involvement. This pattern is more indicative of CJD and

Table 1 Outline of Significant Clinical, Chemical, and Neuroimaging Findings During Hospitalisation

Admission vital signs	• BP: 160/100 mmHg. • HR: 83/minute. • Body temperature: 36.9°C.
Admission neurological examination	• Deep tendon reflexes: Normal in upper and lower limbs. • Babinski and Hoffman signs: Positive bilaterally. • Cranial nerves: Normal bilaterally. • Pupillary and oculocephalic reflexes: Normal. • Rigidity: Present in all four limbs. • GCS score: 9/15 (E4, V1, M4). • Sensory tests, cognitive assessments, and cerebellar examinations could not be performed given the patient's low level of consciousness and generalized rigidity.

(*Continued*)

https://doi.org/10.2147/IMCRJ.S576896

Table 1 (Continued).

Admission work-up	• CBC:
	  ○ Hgb: 11.5 g/dL.
	  ○ MCV: 81.5 fL.
	  ○ WBC: 6900/µL.
	  ○ Granulocytes: 75.5%.
	  ○ Lymphocytes: 1300/µL.
	• Urinalysis:
	  ○ RBC: 3–5/hpf.
	  ○ WBC: 25–30/hpf.
	  ○ pH: 6.
	• ALT: 29 U/L.
	• Albumin: 3.4 g/dL.
	• Creatinine: 0.8 mg/dL.
	• Urea: 43 mg/dL.
	• CRP: 0.9 mg/L.
	• PT: 13.5 seconds.
	• INR: 95%.
	• Total calcium: 9.3 mg/dL.
	• Phosphate: 2.7 mg/dL.
	• Magnesium: 1.83 mg/dL.

Additional investigations	• Regular CSF analysis:
	  ○ RBC: 5/µL.
	  ○ WBC: 0/µL.
	  ○ Protein: 19 mg/dL.
	  ○ Glucose: 51 mg/dL.
	  ○ CRP: 1.3 mg/L.
	• Brain MRI: Bilateral cortical and subcortical hyperintense signals on DWI.
	• EEG: Nonspecific generalized slowing.

Abbreviations: BP, Blood pressure; HR, Heart rate; GCS, Glasgow coma scale; CBC, Complete blood count; Hgb, Hemoglobin; MCV, Mean corpuscular volume; WBC, White blood cell; RBC, Red blood cell; ALT, Alanine transaminase; CRP, C-reactive protein; PT, Prothrombin time; CSF, Cerebrospinal fluid; MRI, Magnetic resonance imaging; DWI, Diffusion-weighted imaging; EEG, Electroencephalography.

excludes other types of rapidly progressive dementia such as autoimmune encephalopathy.[13–15] The symmetrical abnormalities observed on the DWI also indicates a later stage of the disease, aligning with the patient's clinical presentation.[13] Several CJD-mimicking diseases were excluded based on clinical reasoning and biochemical findings Table 2.[16] Real-time quaking-induced conversion (RT-QuIC) and cerebrospinal fluid (CSF) 14.3.3 were unavailable.

Management

Myoclonic jerks and rigidity were managed via Levetiracetam (500 mg/12 hours) and Levodopa/Carbidopa (100/25 mg/8 hours), respectively. The patient also received proper hypertensive therapy and antibiotic therapy (Ceftriaxone 1000mg/24 hours and Levofloxacin 750 mg/24 hours) for urinary tract infection. She was discharged to receive care at home as requested by her family.

Follow-Up

Three months later, follow-up examination revealed a right-sided palmomental reflex and palmar grasp reflex. The patient also exhibited severe cervical dystonia. A few days later, she developed aspiration pneumonia, which led to her death.

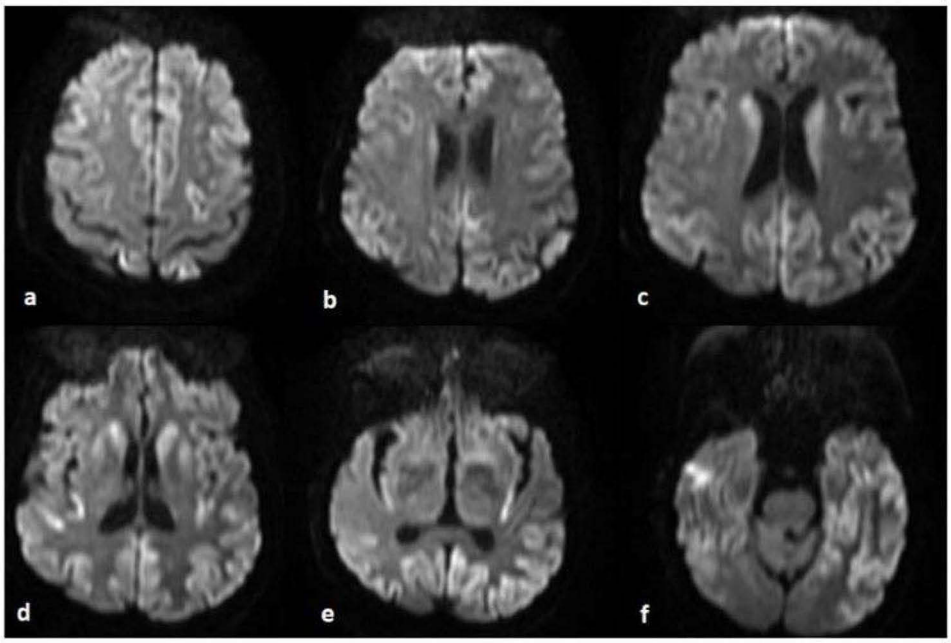

Figure 3 Serial axial diffusion-weighted magnetic resonance images (DWI) demonstrating cortical hyperintensity (cortical ribboning) and basal ganglia involvement. (**a**): Fronto-parietal cortex. (**b–e**): Occipito-temporal cortex. (**f**): Temporal cortex. (**c** and **d**): Note the characteristic high signal intensity in the heads of the caudate nuclei and putamen.

Discussion

The presented case report highlights a probable sCJD in a 72-year-old woman, which to our knowledge, is the first case to be documented in Syria. Given the estimated annual global incidence of 1.5–2 cases per million,[1] the absence of prior reports from Syria, a country with a population of approximately 20 million, suggests a significant surveillance gap and profound underdiagnosis. The case underscores the need for a national prion disease registry to monitor future cases, including iCJD and vCJD, and aid in outbreak prevention. Additionally, the patient's nonspecific symptoms emphasize the importance of considering CJD in differential diagnoses, as delayed diagnosis may stem from not considering prion diseases in the first place.

This report has several limitations, primarily the lack of CSF 14.3.3 testing and advanced diagnostic tools such as RT-QuIC. Consequently, diagnosis relied heavily on clinical examination, exclusion of other conditions, and brain MRI findings. The patient was referred to a private facility to quickly obtain an MRI scan, however, the majority of patients normally face long MRI waitlists in affordable public hospitals, reflecting the broader difficulties of accessing advanced imaging in LMICs.[10] Cultural considerations did not allow for performing post-mortem brain biopsy, which prevented the histological, ie gold standard, confirmation of the diagnosis. Furthermore, as noted in Table 2, the lack of specific ancillary tests means we could not definitively exclude all CJD mimics. However, as detailed in the case discussion, it is unlikely that this case is a CJD mimic as it fits the criteria for a probable CJD diagnosis.

Table 2 Diagnostic Rationale Behind Excluding Various Sporadic Creutzfeldt–Jakob Disease Mimics

Disease	Rationale for Exclusion in Patient
Common neurodegenerative diseases (Alzheimer's Disease, Dementia with Lewy Bodies)	• Rapid progression of signs and symptoms within three months preceding hospitalization. • Absence of cognitive dysfunction prior to the rapid deterioration. • No evidence of focal brain atrophy on MRI.

(*Continued*)

 https://doi.org/10.2147/IMCRJ.S576896

Table 2 (Continued).

Disease	Rationale for Exclusion in Patient
Infection-caused delirium	• Subacute, progressive, and non-fluctuating nature of symptoms. • No improvement after treating urinary tract infection.
Toxic/Metabolic Syndromes[a]	• Normal electrolytes. • No history of alcohol consumption. • Normal hepatic and renal functions.
Immune-mediated encephalitis	• No CSF pleocytosis. • No brainstem abnormality on MRI. • Euthyroid. • No response to five days of intravenous methylprednisolone.
Progressive multifocal encephalopathy	• No history of immunosuppression. • No history of generalized seizures. • No CSF pleocytosis. • No demyelinating lesions in the parieto-occipital region, corpus callosum, thalamus, or basal ganglia.
Subacute sclerosing panencephalitis	• Advanced age. • No known history of childhood measles infection. • No bilaterally synchronous sharp complexes lasting a few seconds on EEG.
Neoplastic, paraneoplastic, and vascular conditions[a]	• No relevant findings on MRI.
Other types of CJD[a]	• vCJD: No history of suspicious food consumption or blood transfusion, and no pulvinar sign on brain MRI. • iCJD: No history of medical or surgical procedures. • gCJD: Negative familial history for any similar cases.

Note: [a] We could not definitively exclude these diagnoses due to the lack of the needed ancillary tests for confirmation.
Abbreviations: MRI, Magnetic resonance imaging; CSF, Cerebrospinal fluid; EEG, Electroencephalography; CJD, Creutzfeldt–Jakob Disease; vCJD, Variant Creutzfeldt–Jakob Disease; iCJD, Iatrogenic Creutzfeldt–Jakob Disease; gCJD, Genetic Creutzfeldt–Jakob Disease.

The economic realities of healthcare in resource-limited countries are another limitation. In such contexts, the economic value of pursuing extensive diagnostic procedures for currently untreatable diseases such as CJD can be questioned, especially when resources are needed for managing treatable conditions. Hence, CJD registries are limited to high income countries, which renders case reports from LMICs essential to better understand the global burden. Collaborative initiatives between research institutions and LMIC healthcare systems could provide cost-effective screening strategies that improve CJD surveillance.

A review of the experiences in other developing nations reveals a consistent challenge in obtaining definitive diagnoses for CJD which limits the comprehensiveness of global epidemiological understanding.[2] Surveillance is often either non-existent or inadequate in many LMICs, contributing to underreporting and frequently relying on clinical criteria rather than pathological confirmation.[2,17] For example, in a series of six cases reported from Peru, all patients were discharged with a probable diagnosis of prion disease because necropsy, the gold standard for definitive confirmation, was not performed.[6] Furthermore, key supportive tests like the 14-3-3 protein assay in CSF were often not performed due to the high cost of extra-institutional private testing.[6] Recent reports from Morocco, Romania, and Saudi Arabia were largely established as probable CJD, relying heavily on clinical criteria, MRI, and sometimes EEG, due to the unavailability or financial constraint of definitive confirmatory tests.[3–5] Similarly, reports from various Asian countries underscore the provisional nature of many diagnoses, often classifying cases as probable or possible sCJD.[17] For instance, Chinese surveillance data, despite being extensive, still categorized the vast majority of sCJD cases reported between 2010 and 2016 as probable or possible rather than definite.[17] In India, a lack of applicable diagnostic techniques meant earlier reports included a significant number of probable cases.[17] Even in more recent Indian case reports, the ability to perform highly specific tests like CSF 14-3-3 and RT-QuIC was

not possible due to financial and logistic constraints.[7] Earlier studies in India (2011–2013) and Kenya (1990–2004) also described numerous probable and possible cases.[8,9] The abundance of probable and possible reports from LMICs reflects similar logistical conditions and underscores the critical need for national surveillance systems in LMICs to track the global burden of prion diseases and improve early detection.

Raising awareness among clinicians about prion diseases is essential to overcome difficulties in diagnosis, mitigate diagnostic delays, and improve case detection rates. Education through workshops, seminars, and accessible resources like clinical guidelines can aid early recognition and enhance understanding and preparedness, ultimately improving diagnosis and patient management in the region.

Conclusion

This case highlights the pressing need to raise awareness of prion diseases among healthcare professionals in resource-limited countries. It underscores the importance of including CJD in the differential diagnosis of rapidly progressive dementia, suggesting the need for prion disease surveillance and registries in LMICs. Despite limitations such as diagnostic resource scarcity and economic constraints, this report serves as a foundational step toward recognizing and addressing prion diseases in Syria and similar regions.

Generative AI Usage

During the preparation of this work, the authors used DeepSeek (DeepSeek latest version model) in order to perform a grammatical check, identify formatting errors, and improve the clarity of the language. After using this tool, the authors reviewed and edited the content as needed and take full responsibility for the content of the publication.

Abbreviations

sCJD, Sporadic Creutzfeldt-Jakob Disease; CJD, Creutzfeldt-Jakob Disease; LMICs, Low- and Middle-Income Countries; gCJD, Genetic Creutzfeldt-Jakob Disease; iCJD, Iatrogenic Creutzfeldt-Jakob Disease; vCJD, Variant Creutzfeldt-Jakob Disease; MRI, Magnetic Resonance Imaging; DWI, Diffusion-Weighted Imaging; FLAIR, Fluid-Attenuated Inversion Recovery; RT-QuIC, Real-Time Quaking-Induced Conversion; CSF, Cerebrospinal Fluid; EEG, Electroencephalography.

Data Sharing Statement

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethics Approval and Informed Consent

Latakia University does not require an ethical approval for the production of case reports. Informed consent was obtained from the patient's next-of-kin for publication of this paper and all accompanying images.

Consent for Publication

Written informed consent for publication was obtained from the patient's next-of-kin. The consenting individual has reviewed and approved the final manuscript and associated materials, including all data, images, and clinical details contained within. They confirm their understanding that the information will be publicly available.

Acknowledgments

The authors would like to extend their gratitude to the patient's family for their support and cooperation.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Funding

This paper did not receive any external funding.

Disclosure

The authors declare no competing interests.

References

1. Ladogana A, Puopolo M, Croes EA, et al. Mortality from Creutzfeldt–Jakob disease and related disorders in Europe, Australia, and Canada. *Neurology*. 2005;64(9):1586–1591. doi:10.1212/01.WNL.0000160117.56690.B2
2. Gao LP, Tian TT, Xiao K, et al. Updated global epidemiology atlas of human prion diseases. *Front Public Health*. 2024;12:1411489. doi:10.3389/fpubh.2024.1411489
3. Guennouni A, Hassar S, Oukassem S, et al. Creutzfeldt-Jakob disease: case report and literature review. *Radiol Case Rep*. 2025;20(5):2577–2580. doi:10.1016/j.radcr.2025.02.007
4. Kelemen K, Kövecsi A, Banias L, et al. A rare case of histopathologically confirmed Creutzfeldt–Jakob Disease from Romania, long route to diagnosis—case report and an overview of the Romanian CJD situation. *J Clin Med*. 2022;11(16):4803. doi:10.3390/jcm11164803
5. Albadr F, Aldusaymani SM, Aldobikhi YA, et al. Creutzfeldt-Jakob disease in a Saudi female: a case report. *Cureus*. 2024. doi:10.7759/cureus.75887
6. Espinoza Ramón SL, Canales D, Calderón C, Díaz D, Barreto-Acevedo E. Enfermedad de Creutzfeldt Jakob esporádica: serie de casos en Perú. *Colomb Medica*. 2024;55(1):e2035821. doi:10.25100/cm.v55i1.5821
7. Pandya KJ, Parmar BK, Pathak JM. Sporadic Creutzfeldt-Jakob disease: a rare case of rapid progressive cognitive decline with special reference to magnetic resonance spectroscopy. *J Assoc Physicians India*. 2024;72(11):e53–5. doi:10.59556/japi.72.0632
8. Hisata Y, Yamashita S, Tago M, et al. Heidenhain variant of sporadic Creutzfeldt-Jakob disease with a variety of visual symptoms: a case report with autopsy study. *Am J Case Rep*. 2023;24. doi:10.12659/AJCR.938654
9. Adam AM, Akuku O. Creutzfeldt–Jakob disease in Kenya. *Trop Med Int Health*. 2005;10(7):710–712. doi:10.1111/j.1365-3156.2005.01435.x
10. Jalloul M, Miranda-Schaeubinger M, Noor AM, et al. MRI scarcity in low- and middle-income countries. *NMR Biomed*. 2023;36(12):e5022. doi:10.1002/nbm.5022
11. Gambetti P, Cali I. Human sporadic prion diseases. In: Zou WQ, Gambetti P, editors. *Prions and Diseases*. Cham: Springer International Publishing;2023:353–373. Accessed March 21, 2025. doi:10.1007/978-3-031-20565-1_18
12. CJD diagnostic Criteria. Creutzfeldt-Jakob Disease (CJD). 2024. Available from: https://www.cdc.gov/creutzfeldt-jakob/hcp/clinical-overview/diagnosis.html. Accessed December 15, 2025.
13. Manara R, Fragiacomo F, Ladogana A, et al. MRI abnormalities in Creutzfeldt–Jakob disease and other rapidly progressive dementia. *J Neurol*. 2024;271(1):300–309. doi:10.1007/s00415-023-11962-1
14. Vitali P, Maccagnano E, Caverzasi E, et al. Diffusion-weighted MRI hyperintensity patterns differentiate CJD from other rapid dementias. *Neurology*. 2011;76(20):1711–1719. doi:10.1212/WNL.0b013e31821a4439
15. Urbach H, Klisch J, Wolf HK, Brechtelsbauer D, Gass S, Solymosi L. MRI in sporadic Creutzfeldt-Jakob disease: correlation with clinical and neuropathological data. *Neuroradiology*. 1998;40(2):65–70. doi:10.1007/s002340050542
16. Mead S, Rudge P. CJD mimics and chameleons. *Pract Neurol*. 2017;17(2):113–121. doi:10.1136/practneurol-2016-001571
17. Rasheed U, Khan S, Khalid M, Noor A, Zafar S. A systemic analysis of Creutzfeldt Jakob disease cases in Asia. *Prion*. 2024;18(1):11–27. doi:10.1080/19336896.2024.2311950